THE KENNEL CLUB
DOG HEALTH

Breed Health and Conservation Plan

Estrela Mountain Dog

Evidence Base

CONTENTS

INTRODUCTION

The Kennel Club launched a new resource for breed clubs and individual breeders – the Breed Health and Conservation Plans (BHCP) project – in September 2016. The purpose of the project is to ensure that all health concerns for a breed are identified through evidence-based criteria, and that breeders are provided with useful information and resources to raise awareness of current health and welfare concerns in their breed, and support them in making balanced breeding decisions.

The Breed Health and Conservation Plans take a complete view of breed health with consideration to the following issues: known inherited conditions, complex conditions (i.e. those involving many genes and environmental effects such as nutrition or exercise levels, for example hip dysplasia), conformational concerns and population genetics.

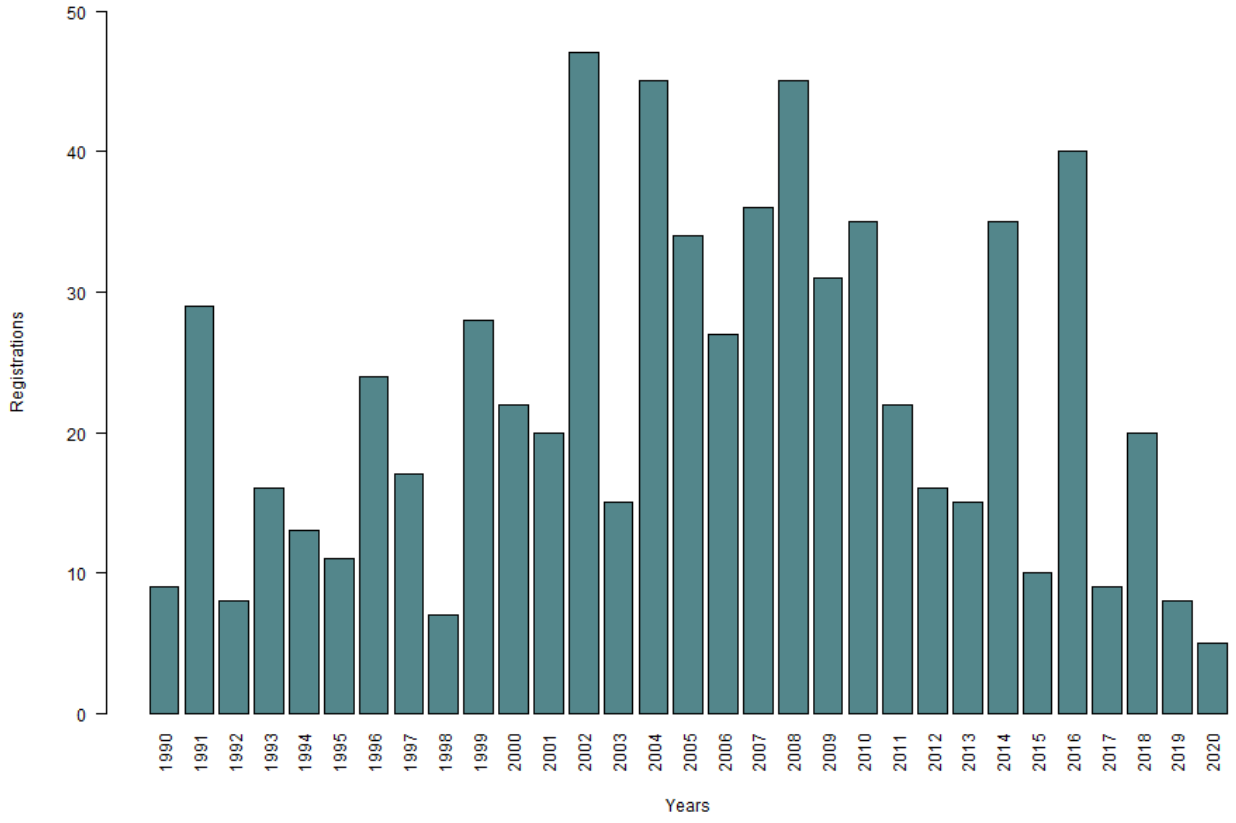
Sources of evidence and data have been collated into an evidence base which gives clear indications of the most significant health conditions in each breed, in terms of prevalence and impact. Once the evidence base document has been produced it is discussed with the relevant Breed Health Co-ordinator and breed health representatives where applicable. Priorities are agreed based on this data and incorporated into a list of actions between the Kennel Club and the breed to tackle these health concerns. These actions are then monitored and reviewed on a regular basis.

DEMOGRAPHICS

The number of Estrela Mountain Dogs registered by year of birth between 1990 and 2019 are shown in Figure 1. The Estrela is a relatively new breed to the UK, with the first dog imported in 1974, and the breed removed from the import register in 1984. The trend of registrations over year of birth (1990-2019) was +0.1 per year (with a 95% confidence interval of -0.4 to +0.6) reflecting the fluctuation in the breed's numbers during this time.

[Put simply, 95% confidence intervals (C.I.s) indicate that we are 95% confident that the true estimate of a parameter lies between the lower and upper number stated.]

Figure 1: Registrations received per year for the Estrela Mountain Dog, between 1990-2020.



BREED HEALTH CO-ORDINATOR ANNUAL HEALTH REPORT

Breed Health Co-ordinators (BHCs) are volunteers nominated by their breed to act as a vital conduit between the Kennel Club and the breed clubs with all matters relating to health.

The BHC's Annual Health Report in both 2018 and 2019, yielded the following response to 'please list and rank the top health and welfare conditions that the breed considers to be currently the most important to deal with in your breed':

1. Hip dysplasia
2. Elbow dysplasia

In terms of what the breed has done to help tackle these listed health and welfare concerns, the breed continues to encourage hip scoring/elbow grading of all breeding stock prior to breeding.

Another priority for the breed in 2019 was to disseminate the results of their health survey and ensure that all the information provided was factual and up-to-date. A summary report of the survey results was emailed to the breed clubs (UK and

abroad) and shared with their members via a newsletter and social media. The BHC has since listed the top health and welfare concerns for the breed as:

1. Orthopaedic issues – in particular hip dysplasia, elbow dysplasia and cruciate ligament rupture
2. Skin problems (including the ear) e.g. eczema and allergies
3. Heart disease – in particular dilated cardiomyopathy (DCM)

In terms of what the breed has done to help tackle these listed health and welfare concerns, the breed has continued to circulate relevant health information from articles and research papers to owners/breeders via their breed club websites, newsletters and social media.

BREED CLUB HEALTH ACTIVITES

The Estrela Mountain Dog has an active Breed Health Coordinator (BHC) and webpages dedicated to health on their club websites which can be found at:

- https://www.estrelamountaindogassociation.com/estrela-health
- https://www.emdc-uk.com/health

A health database for the Estrela Mountain Dog can also be found on Facebook here:

- https://www.facebook.com/groups/149520918506493

BREED SPECIFIC HEALTH SURVEYS

Kennel Club Purebred and Pedigree Dog Health Surveys Results

The Kennel Club Purebred and Pedigree Dog Health Surveys were launched in 2004 and 2014 respectively for all of the recognised breeds at the time, to establish common breed-specific and breed-wide conditions.

2004 Morbidity results: Health information was collected for 23 live Estrela Mountain Dogs of which 8 (35%) were healthy and 15 (65%) had at least one reported health condition. The top categories of diagnosis were dermatologic (25.8%, 8 of 31 reported conditions), musculoskeletal (16.1%, 5 of 31 reported conditions), reproductive (16.1%, 5 of 31 reported conditions), urologic (12.9%, 4 of 31 reported conditions), and aural (6.5%, 2 of 31 reported conditions). The most frequently reported specific conditions were bladder infection/ cystitis (4 cases), hot spot/ pyotraumatic dermatitis/ wet eczema (3 cases), ear infection/ chronic ear infection/ otitis externa (2 cases), failure to push/ uterine inertia (2 cases) and flea allergy (2 cases).

2004 Mortality results: A total of 3 deaths were reported for the Estrela Mountain Dog. The median age at death was 9 years and 9 months (min = 9 years and 9

months, max = 12 years and 10 months). The reported causes of death by organ system or category were combinations, old age and trauma.

2014 Morbidity results: Health information was collected for 24 live Estrela Mountain Dogs of which 14 (58.3%) had no reported conditions and 10 (41.7%) were reported to be affected by at least one condition. The most frequently reported conditions were hip dysplasia (3 cases) and cruciate disease (2 cases).

2014 Mortality results: A total of 1 death was reported for the breed. The age at death for the Estrela Mountain Dog was 8 years. The reported cause of death was a gastric tumour.

Please note: caution should be taken when drawing meaningful conclusions from this data, given the small number of reports for the breed.

Breed-Specific Health Survey 2018

In 2018 the Estrela Mountain Dog Club developed a health survey and received results for 86 dogs. The age of the dogs ranged from 0 – 15 years, with a mean age of 6 years.

The count of conditions by category are shown below, with the most common being bone/ muscle/ joint disorders.

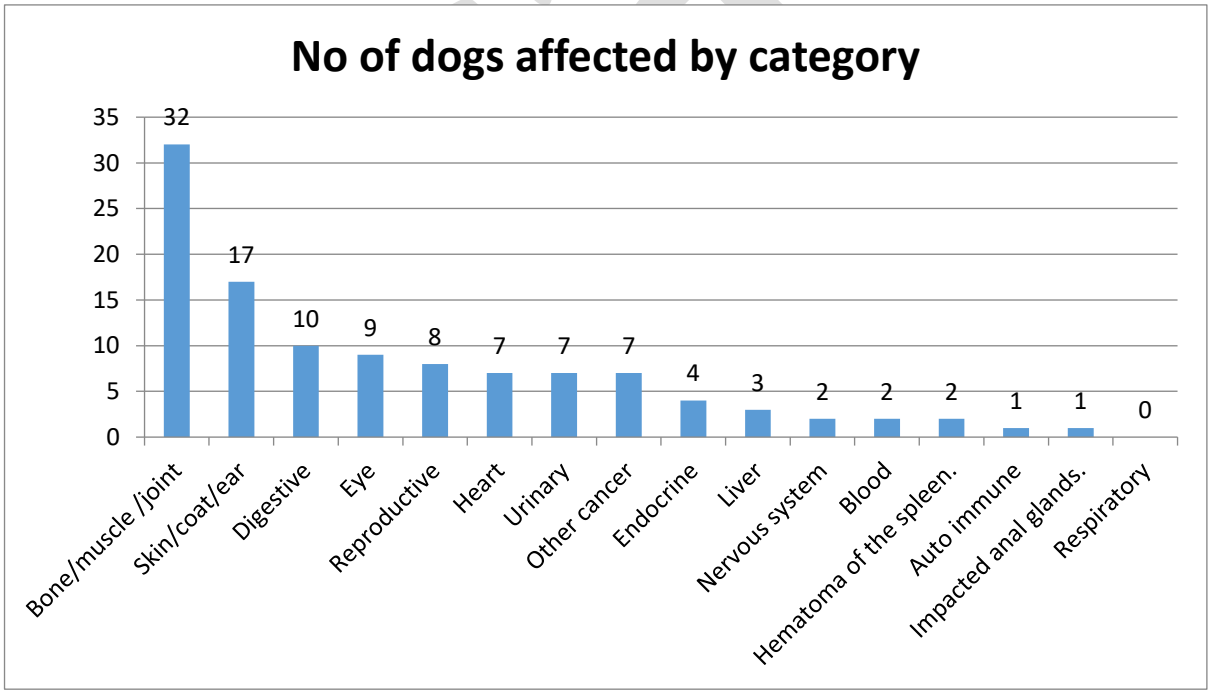


Figure 2: Number of dogs affected per system category in the 2018 breed health survey.

The conditions by count are also given in the graph below, with the top five conditions being hip dysplasia, ear infections/ irritation, suspected food allergies, cruciate ligament disease and arthritis.

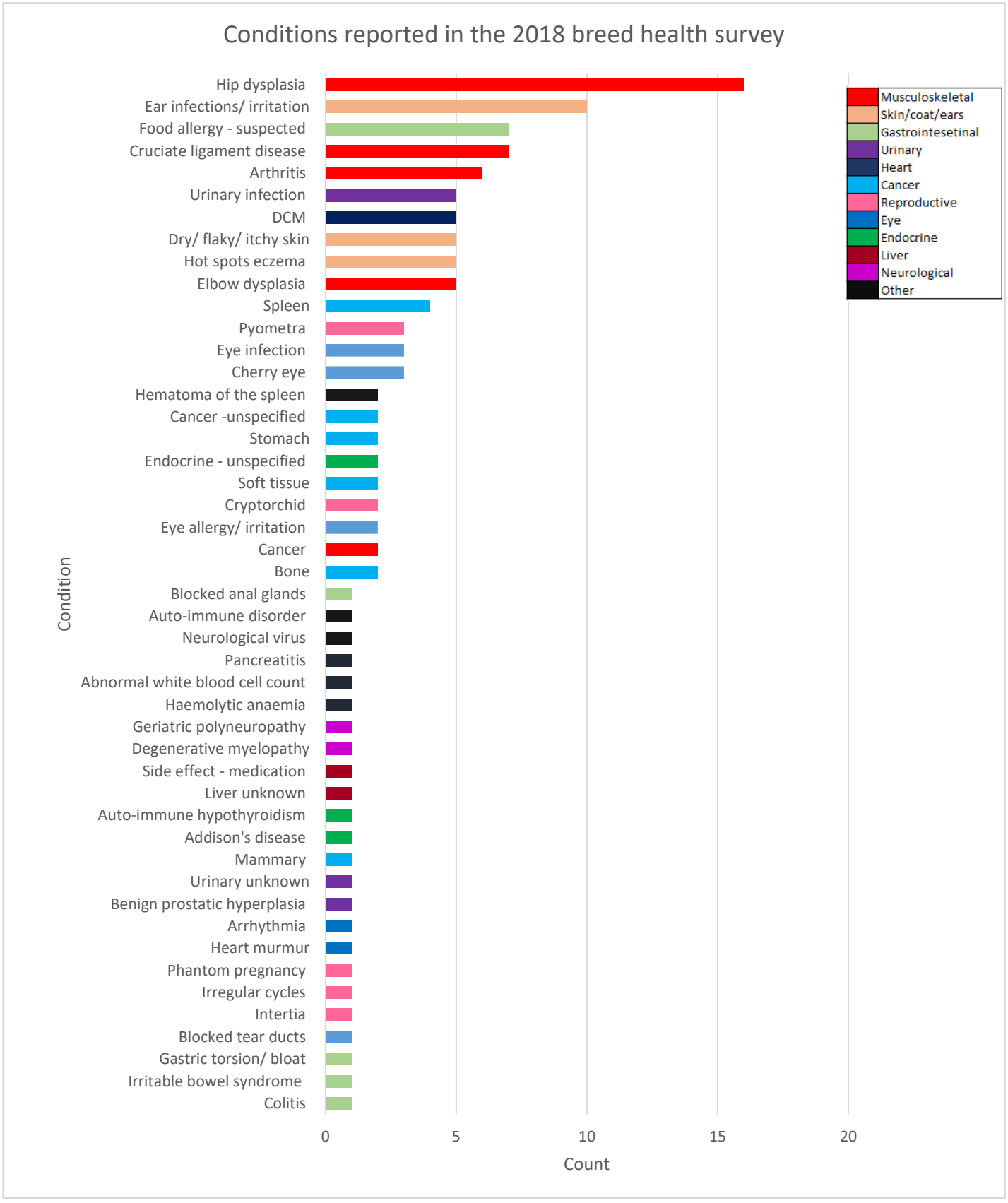


Figure 3: Count of dogs affected per condition in the 2018 breed health survey.

Breed-Specific Health Survey 2019

An Estrela Mountain Dog health survey was disseminated in 2019 and yielded the following morbidity and mortality results:

Health information was collected for 77 live Estrela Mountain Dogs of which 22 (29%) had no reported conditions and 55 (71%) were reported to be affected by at least one condition. The top categories of diagnosis were muscle and joint (38%), skin/ coat/ ear (20%), digestive (12%), eye (11%), and reproductive (11%). The most frequently reported specific conditions were hip dysplasia (16 cases), ear infections (10 cases), cruciate ligament rupture (7 cases), suspected food allergy (7 cases), arthritis (6 cases), dilated cardiomyopathy (5 cases), hot spots (5 cases), and dry or itchy skin (5 cases).

A total of 17 deaths were reported for the Estrela Mountain Dog. The median age at death was 10 years (min = 1 year and 6 months, max = 15 years and 9 months). The most frequently reported causes of death by organ system or category were cancer (47%, 8 of 17), unknown (18%, 3 of 17), and arthritis (12%, 2 of 17). The remaining causes of death had one report each (6%), these were: auto-immune, neurological virus, ruptured spleen and dilated cardiomyopathy.

LITERATURE REVIEW

The literature review lays out the current scientific knowledge relating to the health of the breed. We have attempted to refer primarily to research which has been published in peer-reviewed scientific journals. We have also incorporated literature that was released relatively recently to try to reflect current publications and research relating to the breed.

As the Estrela Mountain Dog is native to Portugal, most publications and research are based on Portuguese dogs. This data is still useful considering the breed's overall population in the UK is relatively small and that it is probable there will be a continuous movement of breeding stock between the UK and abroad.

Musculoskeletal conditions

Hip dysplasia: Hip dysplasia is a complex inherited disorder, influenced by both genetic and environmental factors. As the hip joint deteriorates it can lead to varying degrees of pain, discomfort, stiffness and lameness, and can lead on to the development of osteoarthritis. Hip dysplasia affects a range of breeds, however some breeds, primarily medium and large breeds, have a higher risk of developing the condition.

Estimated Breeding Values (EBVs) remove possible environmental factors, increasing a breeder's ability to predict the genetic (inherited) risk that a dog may pass on to any puppies. A Portuguese study examined the hips and pedigree

information of 215 Estrela Mountain Dogs between May 2002 and December 2006 (Ginja et al, 2008). The results showed stable breeding values between 1991 and 2003 (0.55 to 0.60), and an improvement in 2004 and 2005 (under 0.50). A subsequent study found the prevalence of hip dysplasia in 313 Estrela Mountain Dogs to be 65.8%, confirming the existence and likely predisposition to the condition in the Portuguese Estrela Mountain Dog population (Ginja et al, 2009).

In a more recent study, 525 Estrela Mountain Dogs were radiographically screened and clinically evaluated to determine the prevalence of hip dysplasia in the breed (Martins et al, 2012). The sample was divided into 314 adult dogs (older than twelve months) and 211 young dogs (between four and twelve months). According to the FCI scoring system, 122 adult dogs were graded as normal (FCI grades A or B), and the remaining 192 adult dogs were graded as dysplastic (FCI grades C, D or E). Of the young dogs, 121 were considered to have dysplastic hips.

Elbow dysplasia: Elbow dysplasia is an inherited orthopaedic disorder where the elbow joint does not develop correctly. It is considered to be a complex multifactorial disease with environmental influences. As a dog gets older, the joint undergoes wear and tear and deteriorates, leading to a loss of function. This can cause varying degrees of pain, discomfort, stiffness and lameness, and can also lead on to the development of osteoarthritis or degenerative joint disease.

Alves-Pimenta et al (2013) used the International Elbow Working Group scoring system to evaluate the elbows of 351 Estrela Mountain Dogs. Elbow dysplasia was found in 16.5% (59 of 351) of these dogs, with males (27%, 34 of 127) appearing to have a higher incidence than females (11%, 24 of 224) ($P < 0.05$). The heritability was very low (0.065) and the genetic trend showed a slight positive slope with an improvement in 2004 and 2005. Although the prevalence and heritability of elbow dysplasia in the breed are relatively low, the genetic trends are similar to trends for hip dysplasia, suggesting that the use of selection against hip dysplasia may also result in genetic progress for elbow dysplasia.

Cardiac conditions

Dilated cardiomyopathy (DCM): Dilated cardiomyopathy (DCM) is a heart condition characterized by dilation and reduced contractility of the ventricles resulting in a reduced ability to pump blood around the body. Clinical signs include shortness of breath and lethargy, coughing, weakness, and in severe cases, fainting (syncope) or even death.

An autopsy study used the hearts of 17 Estrela Mountain Dogs (10 from dogs with DCM and 7 from control dogs without DCM) to describe the histologic features of DCM (Lobo et al, 2010). The presence of attenuated wavy fibers (AWFs), fibrosis, and fatty infiltration has been used in different dog breeds for histologic characterization and diagnosis of DCM. There are two histologic types of DCM; fatty infiltration–fibrosis type and AWF type. Usually breeds are associated with having a specific type of DCM, however the findings of this study showed fatty infiltration,

fibrosis, and AWFs coexisted in the myocardium of Estrela Mountain Dogs with DCM. The authors concluded that quantification of fibrosis and observation of AWFs in the left ventricular myocardium are useful in the histologic diagnosis of DCM in Estrela Mountain dogs.

BREED WATCH

The Estrela Mountain Dog is a category one breed, meaning judges are not required to complete mandatory monitoring forms following an appointment as championship certificate level. However, all championship show judges of category one breeds have the opportunity to report on any visible conditions or exaggerations that they consider to be detrimental to the health and welfare of dogs. To date no optional reports have been received for the breed.

The breed would like judges to take extra consideration of incorrect eye conformation or exaggerated head shapes when judging the Estrela Mountain Dog and report any cases of protruding 'third eyelids'.

PERMISSION TO SHOW

As of the 1st January 2020 exhibits for which permission to show (PTS) following surgical intervention has been requested will no longer be published in the Breed Record Supplement and instead will be detailed in BHCPs, and a yearly report will be collated for the BHC. In the past five years, no PTS have been granted for the Estrela Mountain Dog (not including neutering or caesarean sections).

ASSURED BREEDER SCHEME

Currently within the Kennel Club (KC)'s Assured Breeder Scheme there are the following requirements for the Estrela Mountain Dog:

- Hip scoring under the BVA/KC Hip Dysplasia Scheme

There are currently no recommendations for breeding stock of the Estrela Mountain Dog within the KC's Assured Breeder Scheme.

DNA TEST RESULTS

There are currently no recognised DNA tests for the Estrela Mountain Dog.

Whilst DNA tests may be available for the breed, results from these will not be accepted by the Kennel Club until the test has been formally recognised, the process

of which involves collaboration between the breed clubs and the Kennel Club in order to validate the test's accuracy.

CANINE HEALTH SCHEMES

All of the British Veterinary Association (BVA)/Kennel Club (KC) Canine Health Schemes are open to dogs of any breed with a summary given of dogs tested to date below.

HIPS

To date (March 2021), 78 Estrela Mountain Dogs have been hip scored under the BVA/KC Hip Dysplasia Scheme, with a 15-year median hip score of 10 (range 4 - 85) and 5-year of 9 (range 8 - 11).

ELBOWS

To date (March 2021), 15 Estrela Mountain Dogs have been elbow graded under the BVA/KC Elbow Dysplasia Scheme, two dogs being graded 2 in 2007 and 2015, one dog being graded 1 in 2006 and the remaining dogs being graded 0.

EYES

The breed is not currently on the BVA/KC/ISDS Known Inherited Ocular Disease (KIOD) list (formally Schedule A) or Schedule B for any condition under the BVA/KC/International Sheep Dog Society (ISDS) Eye Scheme.

KIOD lists the known inherited eye conditions in the breeds where there is enough scientific information to show that the condition is inherited in the breed, often including the actual mode of inheritance and in some cases even a DNA test.

Schedule B lists those breeds in which the conditions are, at this stage, only suspected of being inherited. As well as the KIOD list and Schedule B, the BVA record any other conditions affecting a dog at the time of examination, which is incorporated into an annual sightings report. Since 2012, just one Estrela Mountain Dog has been eye tested under the BVA/KC/ISDS Eye Scheme and no comments were made.

REPORTED CAESAREAN SECTIONS

When breeders register a litter of puppies, they are asked to indicate whether the litter was delivered (in whole or in part) by caesarean section. In addition, veterinary surgeons are asked to report caesarean sections they perform on Kennel Club registered bitches. The consent of the Kennel Club registered dog owner releases the veterinary surgeon from the professional obligation to maintain confidentiality (vide the Kennel Club General Code of Ethics (2)). Just two c-sections have been reported in the Estrela Mountain Dog to date (2019), one in 2013 and one in 2016.

GENETIC DIVERSITY MEASURES

The effective population size is the number of breeding animals in an idealised, hypothetical population that would be expected to show the same rate of loss of genetic diversity (rate of inbreeding) as the population in question; it can be thought of as the size of the 'gene pool' of the breed. In the population analysis undertaken by the Kennel Club in 2020, an estimated effective population size of **N/A** was reported (estimated using the rate of inbreeding over the period 1990-2019).

Where the rate of inbreeding is negative the effective population size is denoted 'N/A'. This indicates that at this time, the effective population size for the Estrela Mountain Dog is increasing. However, it is important to note that given the small population of the breed this will be constantly fluctuating and breeders should take care to preserve genetic diversity when making breeding decisions.

Annual mean observed inbreeding coefficient (showing loss of genetic diversity) and mean expected inbreeding coefficient (from simulated 'random mating') over the period 1990-2019 are shown in Figure 2. Since 1992, the observed inbreeding coefficient trend has gradually decreased, implying breeders have been carefully selecting mates to restore the diversity in the breed.

It should be noted that, while animals imported from overseas may appear completely unrelated, this is not always the case. Often the pedigree available to the Kennel Club is limited in the number of generations, hampering the ability to detect true, albeit distant, relationships.

For full interpretation see Lewis et al, 2015
https://cgejournal.biomedcentral.com/articles/10.1186/s40575-015-0027-4

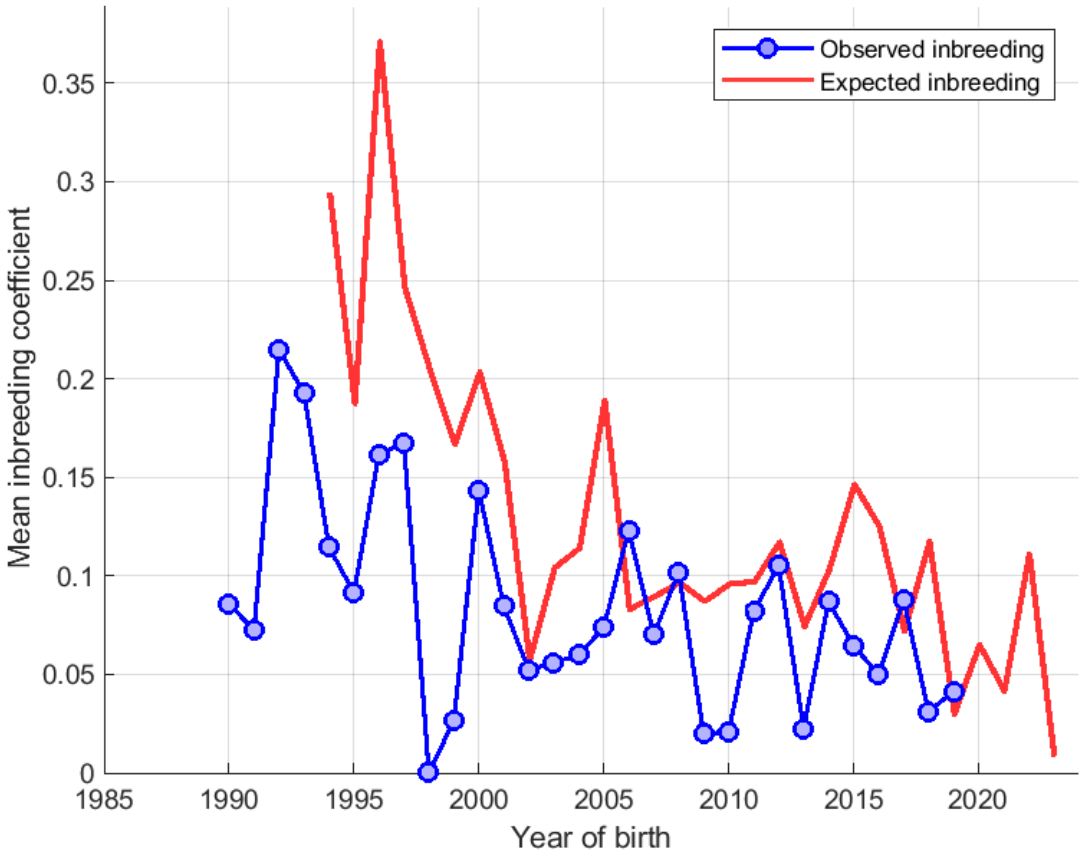


Figure 2: Annual mean observed and expected inbreeding coefficients.

The current breed average inbreeding coefficient for the Estrela Mountain Dog is **4.1%**

Below is a histogram ('tally' distribution) of number of progeny per sire and dam over each of six 5-year blocks (Figure 3). A longer 'tail' on the distribution of progeny per sire is indicative of 'popular sires' (few sires with a very large number of offspring, known to be a major contributor to a high rate of inbreeding). There is evidence of several popular sires being used in the breed, with one sire responsible for over 28% of all registered progeny during 1990-1994. Although this appears to be more under control there is still evidence of several popular sires, however, given the small breeding population of the breed, it is acknowledged that this is difficult to avoid.

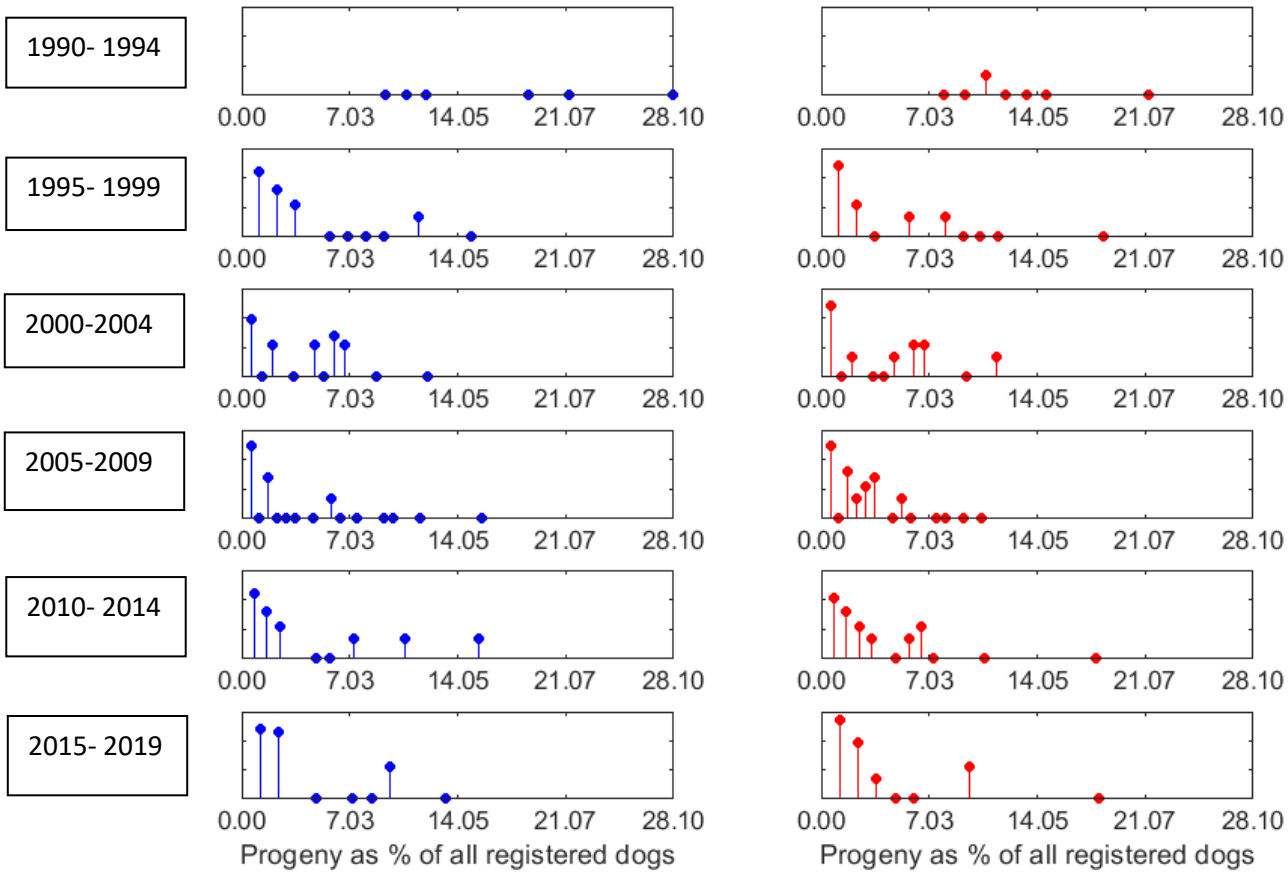


Figure 3: Distribution of the number of progeny per sire (blue) and per dam (red) over 5-year blocks (1990-4 top, 2015-19 bottom). Vertical axis is a logarithmic scale.

CURRENT RESEARCH

The breed are not currently involved in any research.

PRIORITIES

Correspondence between the breed representatives and the Kennel Club was undertaken in April 2021 to discuss the evidence base of the BHCP and agree the priority issues for the health of the breed. The group agreed from the evidence base that the priorities for the Estrela Mountain Dog were:

Priorities:

- Orthopaedic issues – in particular hip dysplasia, elbow dysplasia and cruciate ligament rupture
- Skin problems
- Heart disease – in particular dilated cardiomyopathy (DCM)

At watch:

- Cherry eye
- Progressive retinal atrophy (PRA)

ACTION PLAN

Following the correspondence between the Kennel Club and the breed regarding the evidence base of the Breed Health & Conservation Plans, the following actions were agreed to improve the health of the Estrela Mountain Dog. Both partners are expected to begin to action these points prior to the next review.

Breed Club actions include:

- The Breed Clubs to continue to encourage hip scoring for all breeding stock
- The Breed Clubs to continue circulating health information via their newsletters, websites and social media
- The Breed Clubs to continue monitoring the breed's health via health surveys/breeder reports
- The Breed Clubs to consider the possibility of developing a Health Testing Award Scheme.
- The Breed Clubs to monitor the use of popular sires and raise awareness of the importance of considering genetic diversity when breeding

Kennel Club actions include:

- The Kennel Club to consider adding elbow grading under the BVA/KC Elbow Dysplasia Scheme as a requirement under the Assured Breeder Scheme.
- The Kennel Club to consider adding the following tests/schemes as a recommendation under the Assured Breeder Scheme:
  - Eye testing under the BVA/KC/ISDS Eye Scheme
  - Doppler heart testing
  - Thyroid testing
- The Kennel Club to hold heart testing days and invite breeders of the breed to help in the collation of heart data
- The Kennel Club to produce a piece on the importance of considering genetic diversity and popular sires when breeding, specifically for numerically small breeds

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