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PAPER

Pilot study of head conformation changes over time in the Cavalier King Charles spaniel breed

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Modern interpretation of head conformation in the Cavalier King Charles spaniel (CKCS) has favoured a smaller, more exaggerated, brachycephalic type than originally described in the 1929 breed standard. Recent research studies identified brachycephaly and reduced hind cranium as two conformational (dysmorphic) features that increase risk for symptomatic Chiari-like malformation and secondary syringomyelia (SM). A prospective pilot study investigated the hypothesis that dysmorphic head features could be assessed visually and correlated with risk of SM. Thirteen CKCS, selected from anonymised photographic evidence, were physically appraised by authorised Kennel Club judges using a head shape checklist. These subjective evaluations were then matched with objective measurements of the cranium (cephalic index and rostrocaudal doming) and their subsequent MRI. A positive correlation ($P=0.039$) between the judges' checklist score and rostrocaudal doming (hindskull ratio) and a positive correlation between the cephalic index and hindskull ratio ($P=0.042$) were identified. Five CKCS had no SM and their status tallied with 62 per cent of the judges' evaluation. Although the ability of adjudicators to identify differences in head conformation varied, there was sufficient association between the dysmorphic parameters and the risk of SM to cause concern and propose a larger study in CKCS breed.

Introduction

Chiari-like malformation (CM) and secondary syringomyelia (SM) is an inherited, multifactorial trait.^{1,2} It has been characterised in the Cavalier King Charles spaniel (CKCS) as a complex developmental abnormality of the skull and brain that includes rostrocaudal shortening of the skull base, reduced supraoccipital and interparietal bone, closer proximity of the cervical vertebrae to the skull, and craniocerebral and craniospinal displacement and disproportion.^{3,4} A previous study investigated conformation features and a photographic analysis of the CKCS head shape. Two features were found to be protective against SM; a

decreased cephalic index (less brachycephalic) and more pronounced caudal cranium compared with dogs with SM.⁵ Thus, CKCS with a more exaggerated head type with broader and shorter skulls and increased rostral cranial doming are at increased risk of developing SM. These differing conformational (dysmorphic) features are often described informally as a 'modern' look (exaggerated head type) versus 'traditional' (moderate head type), which is closer to the 'spaniel gentle', that is, the traditional companion spaniel.⁶ Dog breed standards are guidelines which describe the ideal characteristics of a breed both in terms of appearance and temperament and are agreed by breed clubs (not always unanimous internationally). The CKCS Club UK, originated in 1926, describes the breed standard characteristics as having flat skull, shallow stop and ears set high with a 3.8 cm nose.^{7,8} This is illustrated in Foyles Handbook of the CKCS⁹ with the 'right' and the 'wrong' breed features for the CKCS head shape (figure 1). A deep stop and short muzzle (third row) are considered a fault. However, changes in head conformation over the years have led to an 'exaggerated head' with large forward-facing eyes with a steeper stop and higher and shorter head which appeals to the pet buying public.^{10,11} Fifty years later, the CKCS now looks more like the dog depicted as 'wrong' in figure 1, with

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THE BREED STANDARD

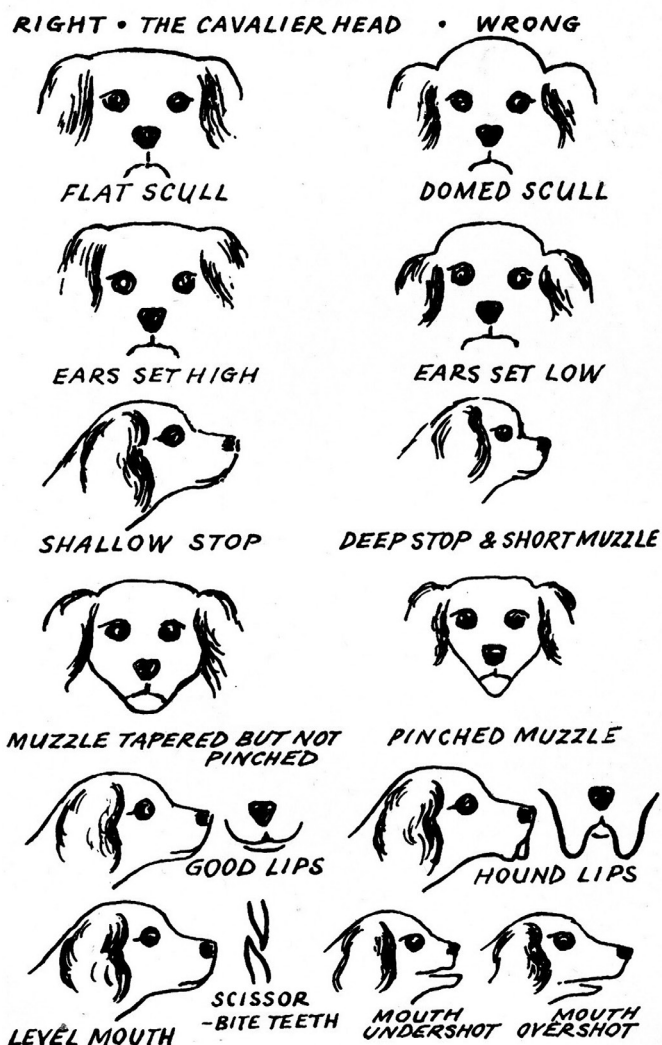


Figure 1 Breed standard as illustrated in Foyles Handbook of the Cavalier King Charles spaniel⁹. Third row indicates the dysmorphic differences in the degree of stop and length of cranium and muzzle. (With permission to publish granted by W & G Foyle). -

respect to pronounced stop with shorter cranium and muzzle length but the breed standard has not altered.

The 'doll like' appearance has been investigated in the Griffon Bruxellois and such features have been shown to be associated with an increased risk and prevalence of CM/SM in the breed^{12 13} which has been linked to a genetic basis for CM/SM.¹⁴ The modern CKCS cranium has been shown to be brachycephalic.¹⁵ Despite this evidence, many breeders have remained sceptical about a relationship with CM/SM and head conformation. In order to resolve the debate between CKCS Club members (particularly in social media) concerning the changes in head shape over time, the Swedish Kennel Club was approached by the authors to fund a prospective pilot study with a minimum of six dogs equally supported by Cavalier Matters Charity (UK).

The subtleties of head are often only appreciated by breed type experts and those who breed and judge at championship level. However, judging dogs is subjective and not an exact science, so this study aimed

to find out if it is possible to identify risk of CM/SM in a dog's head shape through evaluation with the eye alone by breed experts. The advantage of this approach is that it offers an opportunity to deduce the risk of CM/SM by providing an additional tool for breeding selection (without the need for equipment such as calipers) to improve dog welfare and enables both the veterinarian profession and breeders to share their expertise in a unique manner.

Hypothesis

Head conformation (dysmorphic features) is correlated with risk of SM and can be evaluated with the eye alone by breed experts.

Aims

1. Compare two groups of CKCS, identified by breeders to possess different head conformation and establish if one group has increased risk for CM or SM as determined by MRI.
2. Determine if dysmorphic parameters identified in a previous study⁹ are a useful means of predicting MRI diagnosis.

Expected outcomes

This pilot study will:

1. Provide some evidence as to whether recent exaggerated CKCS conformation influences disease and whether further studies should be performed evaluating if this conformation should influence breeding selection.
2. Compare, for the first time, the opinions of adjudicators who are considered expert in the art of assessing dog conformation with an objective, scientific and quantitative MRI assessment.
3. Validate the features of head conformation as predictors of CM and SM status. Currently, CM and SM can only be determined by MRI, which is an expensive test which requires anaesthesia. Determining external conformation features which can be easily measured and which do not require veterinary intervention is an advantage to dog breeders and therefore for dog welfare.

Materials and methods

Selection of study cohort

Six proactive members of the CKCS fraternity (A–F) were invited to the selection panel, representing groups with wide-ranging opinions to ensure no perceived bias or conflict of interest. The remit of the selection panel was to identify the morphological features of the modern and traditional head type, as described in table 1, from photographic submissions and rank each dog into three groups: (1) traditional head; (2) intermediate; (3) modern head, and reject submissions considered unsuitable quality, for example, profile head angle not parallel to the camera or hair coat obscuring shape.

Recruitment

Recruitment of owners who planned diagnostic MRI for health screening purposes was made through social media groups, breed clubs and word of mouth. The selection panel agreed that the terms 'moderate' and 'exaggerated' respectively should be used in the project since this vocabulary was used and understood by the

Table 1 Morphological features of traditional (moderate) and modern (exaggerated) head conformation

Level of characterisation	Head conformation	
	Traditional	Modern type
Primary	A narrower head	A wider head
	A flatter, lower and longer head	Increased doming, that is, a shorter and higher head
	A shallow stop, with less doming of the head towards the front as compared with the back	A deep and pronounced stop, with increased amount of doming towards the front of the head compared with the back
Secondary	A defined occiput	A poorly defined occiput
	Increased distance between nose and eyes	Decreased distance between nose and eyes
	Smaller eyes, better contained within the orbit*	Large, bulging eyes*

*Large and small eyes are terms used by the breeding fraternity and do not reflect globe size, which is constant, but the palpebral aperture so that dogs with 'large eyes' have less orbital covering.

lay community in the context of head conformation. A leaflet/flyer, generated by the selection panel and printed by the Cavalier Matters Charity, widely disseminated to CKCS owners at dog shows, including Crufts and via social media.

Inclusion criteria

Dogs

- ▶ Initially over five years of age because of the late-onset nature of SM but later reduced permitting more recent head conformation breeding selections.
- ▶ Not previously have undertaken diagnostic MRI screening for CM/SM.
- ▶ No medical condition that would prohibit an MRI.

Owners (permission granted by proxy to retain anonymity)

- ▶ Email CKCS photographs of required standard (figure 2) depicting the head from three positions: (1) profile, (2) front, and (3) above.
- ▶ Willing to travel to Swedish/UK veterinary facility in order to participate.
- ▶ Sign a consent and confidentiality statement.

Photographic submissions (figure 2) sent to author LG were anonymised and collated. Profile images of the final selected CKCS cohort were sent to author TJM in order to measure rostrocaudal doming of the cranium with the same grid analysis method as used in the previous retrospective investigation.⁵ This

involved a standard grid made up of four quadrants, aligned with the topline of the muzzle, the lateral border of the first quadrant was aligned with the most rostral point of the cranium and the lateral border of the last, or fourth, quadrant encompassed the remainder, most caudal aspect, of the cranium. Since it is visually easier to compare the hindskull relative to the rest of the cranium rather than as a proportion of the whole cranium, a proportional calculation was made by taking the sum of the three rostral cranial quadrants and divided by the fourth caudal quadrant (ie, the hindskull). This is referred to as the hindskull ratio hereafter.

Physical examination by breed judges

Judges were recruited by invitation via advertising in a local Swedish judges' association and directly via email which provided information about the project. In the UK, requests were made directly to 50 KC registered and experienced Toy Breed judges with specific experience with CKCS. The resulting voluntary judging panel were provided with an information sheet with instructions and checklist based on table 1 (ie, broadness and doming of skull, definition of stop and occiput, and size of eye (palpebral aperture)), and required to sign a consent and confidentiality statement to ensure anonymity throughout the project (online supplementary file S1, Information Sheet). The remit of the judge was to:

1. Physically examine the head conformation of each dog to evaluate the five features on the checklist and generate a score: 1=moderate, 2=intermediate and 3=exaggerated, known hereafter as the checklist score (S). The minimum score is 5, being the most moderate head conformation, the maximum score of 15 the most exaggerated features exhibited.
2. After checklist completion, measure using calipers (Sweden) or oversee (UK) the length (L) and width (W) of the cranium to calculate the cephalic index ($W/L \times 100$). A tape measure was used for dog 13.
3. Confirm the dogs' identity with a microchip scanner, cross-referenced with subsequent MRI.
4. Provide an overall opinion of head conformation by placing the dog into one of two groups: 'moderate' or 'exaggerated'.



Figure 2 Example of photograph submission sent to selection panel by prospective participants.

MRI interpretation by neurologists

After examination, the dogs underwent brain and cervical MRI using a 'reduced cost' screening scheme and according to British Veterinary Association/KC protocol for CM/SM.¹⁶ The MRI DICOM images were sent to author JJ who anonymised them and placed them on a server for download and interpretation by European College of Veterinary Neurology diplomates with recognised expertise in CM/SM, authors CR and HAV.

Statistical analysis

IBM SPSS Statistics V.24 was used for descriptive and statistical analysis. The degree of concordance for the initial selection of study dogs was calculated using the inter-reliability test Kendall W which provides an index value, where 1 is total ranking agreement and 0 is none. Data sets were assessed for normality with Shapiro-Wilk tests and associations between the subjective judicator evaluations and objective variables of cephalic index, hindskull ratio and SM. Spearman's rank correlation coefficient was used with non-parametric variables and Pearson's correlation with parametric variables (significant at 0.05 level (two tailed)). Given the small cohort size further statistical analysis was not considered appropriate.

Results

Subjective evaluation

Out of 67 submitted photographic entries, 66 dogs were accepted by six panellists (A–F) and assigned to three groups: 1=traditional; 2=intermediate; 3=modern (online supplementary file 2). The degree of concordance

for the six panellists was $W=0.663$. Overall, 13 dogs were unanimously selected: eight traditional and five modern. Unfortunately, one of the UK 'modern' dogs died prior to the anticipated MRI and an available dog, designated 'intermediate' by the Panel, was substituted (summary figure 3). Age range of dogs was 2.3–9.2 years (average 5.9 years). 'Traditional' dogs over five years of age (average 6.8 years), 'Modern' dogs average age 4.6 years (substituted dog 3.1 years).

Overall, there were five Swedish KC authorised judges (SWE A–E) and two British (UK A, B). The number of judges available on the day to examine the dogs and provide a checklist score ranged from one to five and checklist scores were averaged when appropriate. All data are provided in online supplementary file 3. There was 95 per cent agreement of placement into the groups (summary figure 3).

Objective evaluation

Measuring for the cephalic index was directed by author LG (SWE) or author TJM (UK) to ensure consistency of methodology. All cranial rostrocaudal quadrant measurements were taken by TJM. All data are provided in online supplementary file 3 and the cephalic index and hindskull ratio in summary figure 1. There was complete concordance of assessment of SM by authors CR and HAV with five dogs clear of SM. Any variations in the measurement of the maximum syrinx transverse diameter by neurologists were averaged (summary figure 3). Dog 13 was reported as having a central canal dilation/presyrinx on the basis of previous research and

Study cohort			Judicators' Subjective Evaluations				Objective Evaluations			
			Photographic	Visual						
Dog code	Age at MRI scan (years)	Country of origin	Panel selection (100% agreement)	KC Authorised Judges grouping	% agreement (actual numbers)	Checklist score 1	Cephalic Index 1	Hindskull ratio ²	ccd/syrinx ³	SM status ⁴
1	9.2	SWE	Traditional	Moderate	100 (4/4)	6.8	89.5	3.96	0	0
2	5.1	SWE	Traditional	Moderate	100 (4/4)	7.2	95.2	3.68	0	0
3	8.6	SWE	Traditional	Exaggerated	100 (4/4)	9.6	95.8	4.45	2mm	2
4	6.1	SWE	Traditional	Moderate	100 (4/4)	6.0	98.9	3.91	0	0
5	6.2	SWE	Traditional	Moderate	75 (4/4)	8.5	96.4	4.17	0	0
6	6.5	SWE	Traditional	Exaggerated	100 (4/4)	12.0	103.1	4.59	4mm	2
7	9.2	SWE	Modern	Moderate	100 (4/4)	7.8	95.5	4.23	4mm	2
8	5.8	UK	Traditional	Moderate	N/A (1/1)	8.0	89.2	3.63	4mm	2
9	8.3	UK	Traditional	Moderate	100 (2/2)	9.0	98.6	4.88	6mm	2
10	7.3	UK	Modern	Moderate	100 (2/2)	8.0	88.6	4.14	0	0
11	1.6	UK	Traditional	Moderate	100 (2/2)	8.5	92.8	3.43	6mm	2
12	3.1	UK	Intermediate	Moderate	100 (2/2)	5.5	84.0	3.71	6mm	2
13	2.3	SLOV	Modern	Exaggerated	100 (2/2)	12.0	88.4	N/A	1mm	1

Figure 3 Tabulated summary of subjective evaluations versus objective measurements. ¹Average if applicable. ²Hindskull ratio=sum total of three rostral quadrants of the cranium; fourth most caudal quadrant (ie hindskull). ³Maximum transverse diameter of (ccd)/syrinx. ⁴SM status 0=unaffected, 1=intermediate, 2=SM affected average if applicable. KC, Kennel Club; N/A, not applicable; SLOV, Slovakia; SM, syringomyelia; SWE, Sweden ccd, central canal dilation.

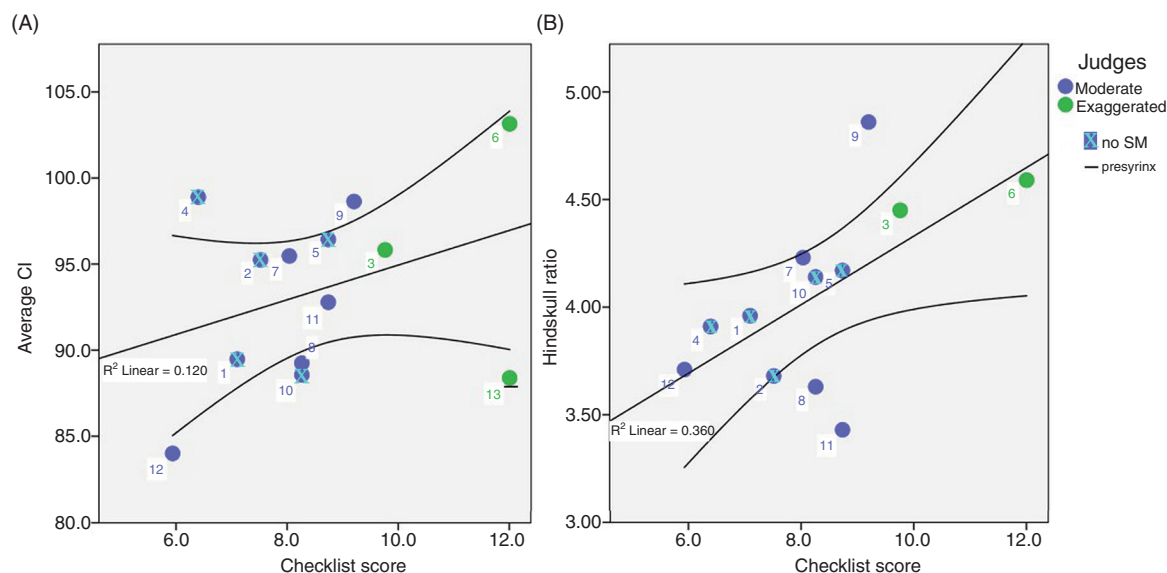


Figure 4 Associations between Judges' checklist score and (A) cephalic index and (B) hinds skull ratio. In graph A, there is a weak positive trend between the checklist score and the average cephalic index (CI). In graph B, there is a significant positive correlation: ($P=0.039$ between the checklist score and the hinds skull ratio, ie, more reduced hinds skull with head shape exaggeration). SM, syringomyelia.

the fact that the dog was 2.3 years of age suggests that this dog is at risk of developing SM later in life.¹⁷

Overarching results

Figure 3 provides details of the study cohort and tabulated to compare the judicators' subjective placement of the dogs into groups with their objective variables.

After confirming variables were normally distributed, the subjective Judge's checklist scores were tested for associations with objective cephalic index and hinds skull ratio (figure 4). The cephalic index was not significantly correlated but indicated a weak positive trend (fitline R^2 linear=0.120, 95% CIs). There was, however, a significant correlation between the checklist score and the hinds skull ratio ($P=0.039$, fitline R^2 linear=0.360, 95% CIs) and a positive correlation between the cephalic index and the hinds skull ratio ($P=0.042$), not illustrated. Dogs without SM, identified with an X, indicate no significant correlation between the checklist score and the MRI results which tallied with 62 per cent of the judges' evaluation. The selection panel had a 75 per cent success rate at matching head shape with SM status (based on photographic evidence), the Swedish judges 86 per cent success rate and the UK judges 20 per cent success rate (figure 3).

Discussion

Breeders have acknowledged that there has been a more brachycephalic interpretation of the breed standard over the last few decades. This pilot study was instigated at the request of the CKCS breed fraternity to investigate the correlation between visually discerned dysmorphic features and risk of CM and/or SM (<https://www.facebook.com/groups/CMSMresearch/>). This prospective investigation demonstrated that it was possible to compare subjective evaluation of head

conformation with objective measurements and revealed a significant correlation between the subjective visual evaluation of head conformation and an objective evaluation of dorsoventral doming using photographs. However, this pilot investigation demonstrated that individual adjudicators can vary in their interpretation of the CKCS breed type (online supplementary files 2 and 3) and also suggests that measuring the cephalic index or rostrocaudal doming alone is not a reliable indicator of brachycephaly but should be taken together with a visual evaluation and take account of other features, such as those on the checklist and the size of the dog.

Figure 5 illustrates four exemplar CKCS in the study with the most extreme range of checklist scores (S) from low to high for Sweden (dogs 1 and 6, top row) and UK (dogs 12 and 9, bottom row). A yellow vertical line indicates the position of the stop and aqua lines and shading indicate the hinds skull. Comparing the dogs with the lowest scores, dog 12 with SM, which appears as an anomaly in figure 4, exhibits greater cranial rostrocaudal doming than dog 1 without SM. Compared with dogs 6 and 9 (both with SM), dog 12 exhibits a longer cranium and muzzle which may have influenced the Judges' subjective decision to classify this dog as 'moderate' (unexaggerated). However, the Panel had previously identified this dog as 'Intermediate'. Specific values for checklist scores and cephalic index should be treated with caution since they are relative to the particular study cohort and do not represent the entire CKCS population.

The concept that increased exaggeration of head shape in the CKCS can be recognised visually and supported by the proven association of brachycephaly with resulting rostrocaudal doming⁵ suggests the possibility for selection against the extreme head shape in the CKCS to enable a reduction in CM/SM incidence.

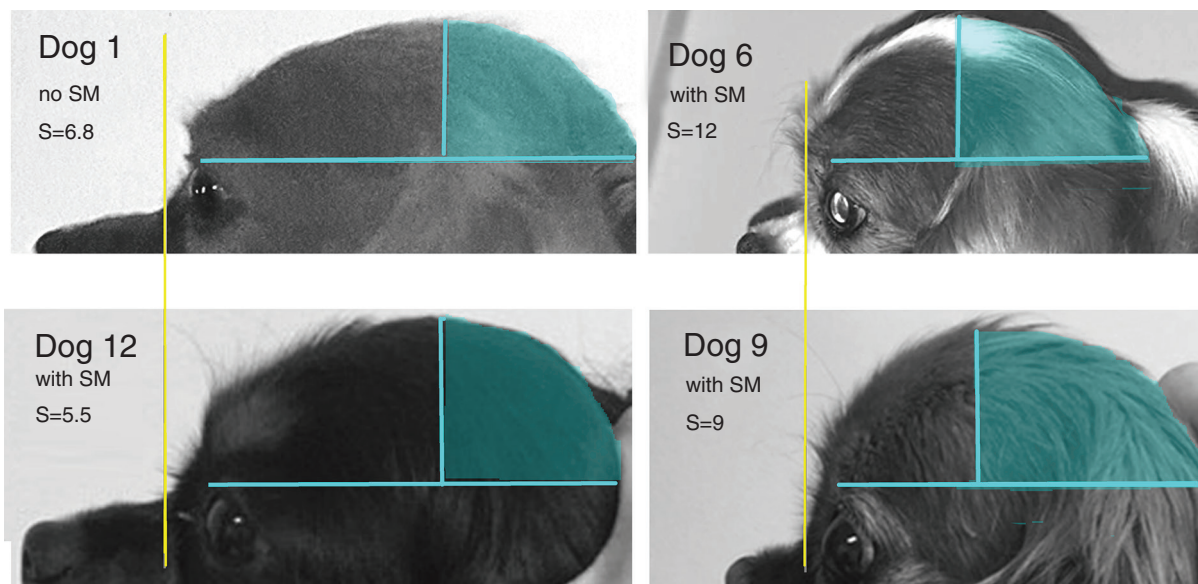


Figure 5 Four Cavalier King Charles spaniel (CKCS) exemplars with the most extreme range of checklist scores (S) in Sweden (top row) and UK (bottom row). Dogs 1 and 12 (lowest checklist scores) are judged to be ‘moderate’. Dogs 6 and 9 (right) have the most exaggerated features. A yellow vertical line indicates the position of the stop and aqua lines and shading indicate the hindskull. Comparing dog 1 without syringomyelia (SM) with dog 12 with SM, the latter has a longer overall cranium and muzzle (proven risk factor). Comparing dog 12 with dogs 6 and 9, also with SM, the former has a longer overall cranium and muzzle.

The dysmorphic features in this pilot study cohort represented the phenotypic extremes available from 66 participants, as unanimously agreed by the selection panellists. Although the terms moderate and exaggerated were useful in describing head conformation, it was apparent that these terms remain subjective and dependant on familiarity with local populations. There may be several reasons for the judges’ discrepancies. Not all individuals had the same opinion as to what was ‘exaggerated’ or ‘modern’ and if one assumes that this phenotype may increase the risk of SM, then further study and defining that phenotype and risk is necessary. The UK judges, who designated ‘moderate’ conformation to all four dogs, may have had prior knowledge of more exaggerated forms or regarded exaggerated type as ‘normal’. None of the judges bred CKCS for the show ring in contrast to the selection panellists. A larger study that involves CKCS breed clubs with their knowledge and experience is recommended.

Limitations of the study

The small number of dogs in the pilot study and the fact that there were less ‘modern-type’ dogs offered for inclusion in the study was a key limiting factor. In practice, this prospective study was far more challenging than envisaged because of the logistical implications and the need for confidentiality and the number of variables out of the control of the investigators. Allowing for average readings, the fact that there was no significant correlation with the checklist score might be possibly a reflection of the small cohort number. However, the integrity of the investigation was enhanced by the involvement and participation of respected CKCS breeders and judges within the CKCS fraternity both in Sweden and the UK. Although the numbers of judges evaluating an individual dog varied, the pilot exercise

demonstrated general agreement, and expected since KC authorised judges are trained to interpret breeding standards consistently.

The quality of the photographs illustrating the head to reveal dysmorphic features was possibly a limiting factor in the initial selection of participants and in taking cranial measurements investigating rostral-caudal doming. The occiput is not always prominent in the CKCS and the angle of the head with foreshortening can be deceptive but the unanimity of the panellists mitigated this. Although the measurements taken from photographs of the head profile were performed by a single skilled veterinary researcher, placement of the grid was subjective.

Age may have been a limiting factor since many breeders do not MRI scan their older dogs, meaning they were not available for potential recruitment into the study. It is quite probable that regional variations in popularity in head conformation might exist between countries such as Sweden and the UK, and lead to a bias in entries. However, investigating the subjective nature of evaluating head conformation was a primary aim for this project.

Potential impact of study on breeding selection

The study identified 7/12 dogs with syrinx of 2–6 mm (figure 3) and confirms the prevalence of SM in the breed,^{17–20} and that, in proportion to the rest of the cranium, the hindskull is reduced with increased cephalic index (figures 1 and 2). Since the reduced caudal skull had already been proven^{12,21} and purported to be recognised by some breed experts, it might provide greater guidance in judging criteria to avoid conformation extremes in breeding. Although the study focuses on the extremes of conformation, over half of the participants entered into the study based on

photos were grouped as ‘intermediate’ by the panellists. Further investigation using a more detailed checklist might highlight additional features which could provide breeders with more information about protective and risk conformation.

The role of breeders and pet owners in selection of dogs is as crucial in ensuring dog welfare as input by the veterinarian profession.²² Recent publications^{11 23} confirm that the pet buyers are greatly influenced by appearance and breeders respond to this by providing the market. There is therefore an important role to play in education of the public, especially children who influence their parents or the media who use extreme phenotypes in advertising, and a larger study might be helpful in providing evidence. It should be advantageous for breeders to select for a pleasing appearance of the CKCS breed but avoid the extremes that increase risk of such conditions as CM/SM. It has been demonstrated that head conformation can be modified,^{24 25} therefore with careful selection and the possibility of outcrossing it may be possible to improve the conformation of the CKCS if necessary.

Conclusion

This study investigated, for the first time, the opinions of adjudicators who are experts in the art of assessing dog conformation with quantitative assessment. It explored the concept that the head conformation of CKCS had evolved over the hundred years since first registered with the KC with the use of the vocabulary for describing the head shape into two groups as ‘traditional versus modern-type’ by panellists and ‘moderate versus exaggerated’ by Toy Breed judges. The hypothesis that head conformation (dysmorphic features) could be assessed visually was shown to be possible but any correlation with risk of SM in the CKCS was inconclusive in this small sample study. The results suggest there was sufficient association between the judicators’ assessment and the risk of SM to propose a larger study.

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Competing interests None declared.

Ethics approval NASPA, a subcommittee of the Animal Welfare and Ethical Review Board, approved this study (reference NASP-2015-009-SVM).

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