

## Title

Is a fashionable and modern head shape an inadvertent risk for Syringomyelia (SM)? - investigation of head conformation in the Cavalier King Charles Spaniel.

## Brief outline

Judging dogs is an art not a science. The subtleties of head conformation are often only appreciated by breed type experts and in particular those who breed and judge at championship level. In this study breeders will have the opportunity to prove or disprove a hypothesis which has been debated passionately for a decade. This hypothesis centers on whether head shape increases the risk for the painful disease syringomyelia and if this is the case, whether such a head shape can be identified by breed experts and without the need for measurements. The study will compare 6 dogs with 'moderate head' (described as having a relatively shallow stop and longer lower head) with 6 dogs with an 'exaggerated head' (described as having a steeper stop, higher and shorter head). The 12 dogs will be selected by consensus agreement by an expert panel of UK and Swedish judges. The judge selection will be made by mutual agreement between all stakeholders to ensure there is no perceived bias. The Chiari-like malformation (CM) and syringomyelia (SM) status will be determined by magnetic resonance imaging (MRI) of the brain and cervical spinal cord. The MRI DICOM will be graded by diplomate neurological with specialist knowledge of CM and SM who are blinded to the conformation groups. In addition to this subjective assessment the dogs' head conformation and MRI will be objectively measured using parameters determined in recent studies [1, 2].

## Background

Syringomyelia (SM) is characterised by fluid-filled cavitation within the spinal cord. This is most commonly associated with Chiari-like Malformation (CM), which is a malformation resulting in a mismatch between skull and brain volume and overcrowding of the foramen magnum [3-5]. Compression of the cerebrospinal fluid channels through the foramen magnum is associated with development of syringomyelia with associated pain and neurological deficits, including paresis, ataxia and scoliosis. This condition may develop progressively over time. SM is ubiquitous Cavalier King Charles spaniels (CKCS) [6, 7] with a high heritability [8].

In humans, some Chiari malformation phenotypes are associated with classic facial and skull features, for example Chiari malformation secondary to craniosynostosis (premature skull suture fusion). There is a suggestion that dysmorphology may also be useful for dogs. A recent paper identified that Griffon Bruxellois dogs with CM and SM had a shortened skull base, which was compensated for by an increased height of the head [2]. An earlier study of radiographs in the Griffon also identified changes in skull shape [9]. Increased forehead height or "bombe" is considered by some to be a desirable feature and there is concern that breeders may be inadvertently selecting for a trait with might predispose to disease. The head and skull shape changes from normal to a CM and SM affected Griffon Bruxellois, described in [2, 9] are illustrated in the following videos:

Changes in skull and brain: <http://youtu.be/qLIufwaxtVQ>

Profile of head: <http://youtu.be/O7IUTTKE0ro>

Front view of head: <http://youtu.be/qWPur5EephI>

A recent study determined if any conformational features were associated with CM and SM in the CKCS. Two head shape features were found to be protective against syringomyelia and the study suggested that a more exaggerated head type may increase risk of disease [1]. There has been much debate about the differences in head shape associated with SM in the CKCS. A dedicated social media page (CMSM Research, which has over 1,000 members) was established set up to encourage discussion and collaboration between breeders, researchers and pet owners. This lively forum has stimulated interest from breeders around the world and breed experts have highlighted the differences between more exaggerated and more moderate head types (Table 1). These conformational features are often described informally as a “modern” look (exaggerated head type) versus “traditional” (moderate head type), which is closer to the “spaniel gentle”, i.e. the traditional companion spaniel [10].

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

Level of characterisation	Modern head type	Traditional head type
<i>Primary characteristics</i>	a wider head	a more narrow head
	increased doming, i.e. a shorter and higher head	a flatter, lower and longer head
	a deep and pronounced stop, with increased amount of doming towards the front of the head as compared to the back	a shallow stop, with less doming of the head towards the front as compared to the back
<i>Secondary characteristics</i>	a poorly defined occiput	a defined occiput
	decreased distance between nose and eyes	increased distance between nose and eyes
	large, bulging eyes	smaller eyes, better contained within the orbit

Figure 1. Variety of head shapes of affected and unaffected CKCS discussed in the social media group.

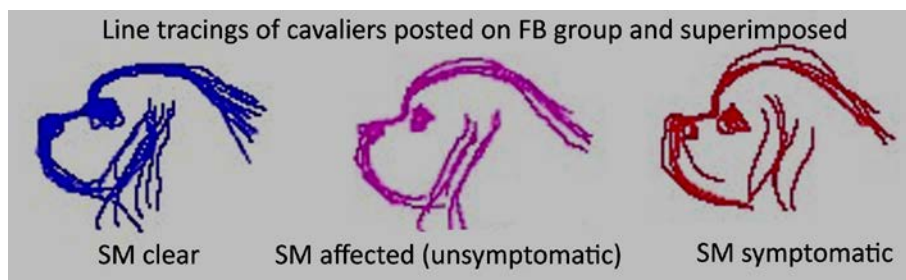


Figure 2. Range of head shapes in the cavalier breed from more exaggerated to more moderate examples.



During the forum debate, geneticist and member of the CMSM Research social media group, Bruce Cattanaach suggested that six CKCS with moderate heads and six CKCS with exaggerated heads should have brain and cervical MRI and be CMSM graded in a blind trial. The dogs should all be over five years old and the MRIs would be performed free of charge to the owners. Dr Cattanaach wrote: "Judging dogs is not an exact science. The subtleties of the head can only really be seen by people in the breed and by those who breed and judge these dogs at championship level. They are the people who need to be involved – with their expertise and their ‘top’ dogs."

These comments and the subsequent debate inspired this study.

### **Hypothesis**

Head conformation (dysmorphic features) is not correlated with risk of CM and / or SM

### **Aims**

1. Compare two groups of CKCS with 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 study [1] are a useful means of predicting MRI diagnosis.

### **Material and Methods**

A judging panel (15 individuals) will be established. To ensure that there is no conflict of interest, this group is likely to be respected toy dog judges outwith the CKCS community but selected by stakeholders within the CKCS. Dogs from Sweden and UK will be proffered for selection from a wider pool of willing owners in possession of dogs over 5 year old. The judging panel will select six CKCS that they deem to have a ‘traditional head’ and six dogs with “modern head” (according to the characteristics outlined above). The judges will make their selection based on front and profile photographs of the dogs and must be in unanimous agreement.

The twelve dogs will undergo brain and cervical MRI using a ‘reduced cost’ screening scheme, which obtains a limited MRI (3 sequences) to determine CM and cranial cervical SM status. The economic cost of the MRI is similar in UK and Sweden. A minimum of 6 dogs will be imaged in Sweden.

The MRI DICOM will be placed on a server for download and interpretation by diplomates of the European College of Veterinary Neurology that have recognised expertise in CM and SM. These diplomats are

1. Dr. Clare Rusbridge BVMS DipECVN PhD MRCVS  
Chief of Neurology Fitzpatrick Referrals and Reader in Veterinary Neurology  
University of Surrey, School of Veterinary Medicine Faculty of Health and  
Medical Sciences
2. Prof. Holger Volk, DVM PhD DipECVN FHEA MRCVS.  
Clinical Director of the Queen Mother Hospital, Head of the Neurology &  
Neurosurgery service and Professor of Veterinary Neurology and  
Neurosurgery, Royal Veterinary College.

The diplomates will be blinded to the dogs' head shape group. Reports on CM and SM status will be generated for the owners of the scanned dogs. These reports will be based on the criteria used by British Veterinary Association / Kennel Club CMSM health scheme [11]. Variations of this scheme are used in other countries including Netherlands, Finland and Australia. In addition, other features deemed useful by breeders will be reported e.g. degree of ventriculomegaly and presence of otitis media with effusion (also referred to as primary secretory otitis media).

In addition, a quantitative MRI analysis will be undertaken by Surrey University PhD student Penny Knowler using a protocol established previously for the Griffon Bruxellois [2] and a quantitative conformation analysis will be made or supervised by Bristol Veterinary student Thomas Mitchell using a protocol established previously for the a conformational indicators study [1]. Both researchers will be blinded to the dogs' head shape group and the MRI results.

### **Expected outcomes.**

1. CKCS breeders have been accused that CM and SM are a consequence of "bad breeding" and "unnatural head shape selection". However, no objective evidence exists for this accusation and, more importantly, there is no guidance as what head shape reduces risk of disease. This study will provide some evidence as to whether modern conformation influences disease and whether this conformation should influence breeding selection. Alternatively in the event that the above hypothesis is proved then breeders can be more confident that their choice of selecting for a modern head shape will not increase likelihood of SM.
2. This study will, for the first time, compare the opinions of adjudicators that are expert in the art of assessing dog conformation with an objective, scientific and quantitative MRI assessment.
3. This study will 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 anesthesia. Determining external conformation features which can be easily measured and which do not require veterinary intervention is an advantage to dog breeders.

## References

1. Mitchell TJ, Knowler SP, van den Berg H, Sykes J, Rusbridge C. Syringomyelia: Determining Risk and Protective Factors in the Conformation of the Cavalier King Charles Spaniel Dog. *Canine Genetics and Epidemiology*. 2014;submitted under review
2. Knowler SP, McFadyen AK, Freeman C, Kent M, Platt SR, Kibar Z et al. Quantitative analysis of Chiari-like malformation and syringomyelia in the griffon bruxellois dog. *PloS one*. 2014;9(2):e88120. doi:10.1371/journal.pone.0088120.
3. Rusbridge C. Chiari-like malformation and syringomyelia. *European Journal Companion Animal Practice* 2013;23(3):70-89. doi:<http://ejcap.fecava.org/#/en/241046/109421/cover-ejcap-online-233.html>.
4. Rusbridge C, Greitz D, Iskandar BJ. Syringomyelia: current concepts in pathogenesis, diagnosis, and treatment. *Journal of veterinary internal medicine / American College of Veterinary Internal Medicine*. 2006;20(3):469-79.
5. Cross HR, Cappello R, Rusbridge C. Comparison of cerebral cranium volumes between cavalier King Charles spaniels with Chiari-like malformation, small breed dogs and Labradors. *The Journal of small animal practice*. 2009;50(8):399-405. doi:10.1111/j.1748-5827.2009.00799.x.
6. Rusbridge C, Knowler SP. Inheritance of occipital bone hypoplasia (Chiari type I malformation) in Cavalier King Charles Spaniels. *Journal of veterinary internal medicine / American College of Veterinary Internal Medicine*. 2004;18(5):673-8.
7. Parker JE, Knowler SP, Rusbridge C, Noorman E, Jeffery ND. Prevalence of asymptomatic syringomyelia in Cavalier King Charles spaniels. *The Veterinary record*. 2011;168(25):667. doi:10.1136/vr.d1726.
8. Lewis T, Rusbridge C, Knowler P, Blott S, Woolliams JA. Heritability of syringomyelia in Cavalier King Charles spaniels. *Vet J*. 2010;183(3):345-7. doi:10.1016/j.tvjl.2009.10.022.
9. Rusbridge C, Knowler SP, Pieterse L, McFadyen AK. Chiari-like malformation in the Griffon Bruxellois. *The Journal of small animal practice*. 2009;50(8):386-93. doi:10.1111/j.1748-5827.2009.00744.x.
10. Casius J. *Gesner de canibus Anglicus (Englische Dogges)* 170, Strand, WC: The Bazaar Office; 1576.
11. BVA. Chiari Malformation/Syringomyelia Scheme (CM/SM Scheme). In: *Canine Health Schemes British Veterinary Association* [http://www.bva.co.uk/canine\\_health\\_schemes/ChiariMalformationSyringomyeliaSchemeCMSMScheme.aspx](http://www.bva.co.uk/canine_health_schemes/ChiariMalformationSyringomyeliaSchemeCMSMScheme.aspx). 2013. Accessed 8th July 2013.