Prevalence of Chiari-like malformation and Syringomyelia in a number of look-alike toy breeds in the Netherlands

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Introduction

In 2007 a Working Group round table defined Chiari-like malformation (CM) as "a decreased volume of the caudal fossa and a protrusion of the cerebellum through the foramen magnum"(2). A more recent research article reports CM is caused not only by the abnormally small caudal fossa but also by the increased cerebellar volume (3).

Syringomyelia (SM) is the development of cavities filled with extracellular fluid, syringes, in the central canal of the spinal cord. They usually originate in the white substance and can extend into the dorsal horn (2,4). It is generally appreciated that SM can be caused by various afflictions such as trauma, tumors and CM. Within the context of Chiari-like malformation it is referred to as chiari-associated syringomyelia. Multiple hypotheses have been suggested as to why CM can cause SM in the last 70 years (5). One of the more recent theories considers the possibility of the syrinx being filled with extracellular fluid, instead of CSF, caused by mechanical distension (1). The intramedullary pulse pressure theory suggests de increased pressure in front of the obstruction and the decreased pressure caudally causes a distension in the latter region (1).

The most common clinical sign that is seen in CM/SM is pain which can manifest itself in different ways. Hyperesthesia in the cervical region can cause an intolerance to wearing a collar and "phantom scratching" is regularly observed. Some patients can cry out in pain with no apparent reason and have an exercise intolerance (1,6). Though it is generally thought only dogs with SM are symptomatic, there have been reports of patients with only CM exhibiting signs. Coexisting conditions of the craniocervical junctions can intensify the clinical signs (1).

The prevalence of CM in the Cavalier King Charles Spaniel population is approximately 95% but dogs of other toy breeds like the Griffon Bruxellois and the Chihuahua can also suffer from this disease (6). A recent study reports a prevalence of 65% CM and 52% SM in the American Griffon Bruxellois population (7).

MRI is the diagnostic tool for diagnosing CM/SM. Herniation of the cerebellum into the foramen magnum is visible but may not reflect the severity of the clinical signs. Syringes may show up and the maximum width can be associated with the clinical signs. If a syrinx isn't seen on the MRI it is still possible that one is located in another part of the central canal. Abnormalities are most severe in the upper cervical and thoracic segments (4,5).

Materials and Method

MRI scans

MRI's are provided by the Companion Animal Clinic Den Heuvel (city of Best, The Netherlands) and only scans of dogs without a pedigree were selected. A total of 262 scans were contributed of which 32 proved to be pedigree dogs based on the microchip number. Another 48 scans were excluded as a consequence of the quality of the scan and therefore the interpretability. The name and address, the chip number, look-a-like breed, the sex and the age of the dog at the moment of scanning was documented.

Grading system

The scans are being scored according to the BVA/KC scheme. It makes use of three age groups. Group A consists of dogs over 5 years of age, dogs in group B are between 3 and 5 years old and group C is younger than 3 years. The CM is graded in either 0 (no CM), 1 (indentation of the cerebellum, but visible CSF between cerebellum and brain stem, or 2 (herniation of the cerebellum through the foramen magnum, no visible CSF). SM is also graded in 0 (no SM), 1 (presence of a syrinx with a diameter smaller than 2 mm), 2 (presence of a syrinx with a diameter larger than 2 mm).

Results

As mentioned, after selection, a total of 182 scans remained. The largest group is the non-pedigree Cavalier King Charles Spaniel, which consists of 152 patients. There are 11 Chihuahua's, 5 French Bulldogs, 1 Jack Russel Terrier, 1 King Charles Spaniel, 1 Border Terrier, 1 Lhasa Apso, 3 Maltese, 2 Pugs, 3 Shih Tzu's and 2 Yorkshire Terriers. All without a pedigree and as such named look-alikes. The youngest dog was 0,4 years old, the oldest 14,1.

Look-a-likes	Number	Median	Range
Cavalier King Charles Spaniel	152	4	0,4 – 9,9
Chihuahua	11	2,6	1 – 7,5
French Bulldog	5	2,2	1,7 – 5,4
Jack Russel Terrier	1	6,3	x
King Charles Spaniel	1	0,8	x
Border Terrier	1	0,7	X
Lhasa Apso	1	4	X
Maltese dog	3	7,6	3,7 – 14,1
Pug	2	8,2	7,6 – 8,7
Shih Tzu	3	5,4	4,1 – 8,4
Yorkshire Terrier	2	4,6	2,1 – 7,1

Table 1: number of dogs, median and range in years.

Cavalier King Charles Spaniel look a-likes

The largest group, the look-alike Cavalier King Charles Spaniel, consisted of 51 dogs with CM, 3 dogs with SM, 95 dogs with CM/SM and only 3 with no CM or SM.

CKS	CM	SM	CM/SM	No CM/SM
A	15	0	41	1
B	14	3	28	2
C	22	0	26	0
Total	51 (33,5%)	3 (2%)	95 (62,5%)	3 (2%)

Table 2: number and percentage of CM and SM in the Cavalier King Charles Spaniel in the three age groups.
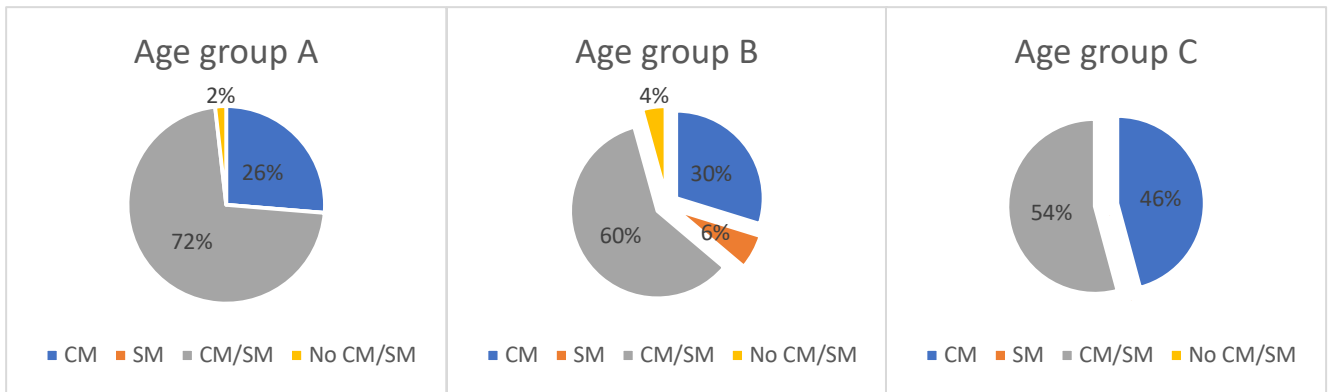
Fig 1: percentage of CM and SM in the Cavalier King Charles Spaniel in the three age groups.

Chihuahua look a-likes

2 Chihuahua's had CM as well as SM, 3 had CM, 3 had SM and 3 had no CM or SM.

Chihuahua	CM	SM	CM/SM	No CM/SM
A	0	1	1	0
B	1	1	0	0
C	2	1	1	3
Total	3 (27,3%)	3 (27,3%)	2 (18,1%)	3 (27,3%)

Table 3: number and percentage of CM and SM in the Chihuahua in the three age groups.
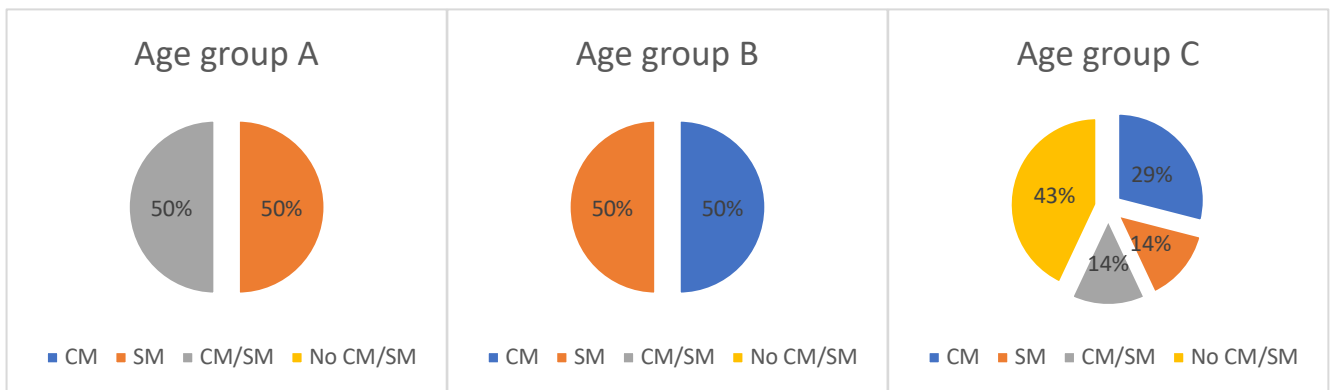

Fig 2: percentage of CM and SM in the Chihuahua in the three age groups.

Other toybreed look a-likes

Neither the Border Terrier, the Lhasa Apso or the Pugs could be diagnosed with CM and/or SM. Because of the small number of scans of the French Bulldog, the King Charles Spaniel, the Maltese dog, the Shih Tzu, the Yorkshire Terrier and the Jack Russel Terrier, the results are compiled into one table.

Look-a-likes	CM	SM	CM and SM	No CM and/or SM
French Bulldog	2	2	1	0
King Charles Spaniel	0	0	1	0
Maltese Dog	1	0	2	0
Shih Tzu	1	0	0	2
Yorkshire Terrier	2	0	0	0
Jack Russel Terrier	0	0	1	0

Table 4: number of CM and SM in the French Bulldog, King Charles Spaniel, Maltese Dog, Shih Tzu, Yorkshire Terrier and the Jack Russel Terrier.

Discussion

It is important to realize this study is not an incidence study. The aim of this retrospective study was to examine the prevalence of CM/SM in toy breed look-alikes. Due to the lack of screening programs in look-alike breeds, most likely all of the scans were made for clinical reasons. There is no information available concerning the incentives for the scans.

The biggest concern of this study is the number of scans available. Only the Cavalier King Charles look-alike group is large enough for statistical evaluation There are multiple reasons for this lack of data. For instance people with look-a-like dog might not be well informed on breed specific conditions and might therefore not have their dog checked. A second possibility is they are less concerned with breeding so they are not interested in scanning for hereditary ailments. Lastly one must keep in mind the economic argument. An MRI is not an inexpensive diagnostic tool and so many may refrain from utilizing it. These reasons are also possible biased and have most likely influenced the representation of the look-alike population.

Other retrospective studies have produced percentages of CM affected dogs up to 95% prevalence in Cavalier King Charles Spaniels. This complies with the 96% prevalence that is found in this study. There seems to be no age significance in grading for CM, but for SM, as a complication of CM, there is. A research article found that the percentage of SM increased with age (3), as seems to be the case with the Cavalier King Charles Spaniel look-alikes. Of age group C, 54% could be diagnosed with SM. This is in contrast with the 32% found in the pedigree CKCS. A plausible explanation for this difference is the presence of the scanning program. As discussed, most likely all our scans were made because of clinical signs the dogs were showing. Around 60% of age group B and 54% of age group A have SM as well as CM, which is more similar to the 54% and 60% percentages found in the pedigree dogs.

The only conclusion we can make based on the data of the other look-alikes is that CM does occur in Shih Tzu and Yorkshire Terrier look-alikes and CM/SM can develop in Chihuahua, French Bulldog, King Charles Spaniel, Maltese dog and Jack Russel Terrier look-a-likes. Unfortunately, we cannot make any statements considering the prevalence. Due to lack of data we cannot make any assumptions considering the appearance of CM/SM in the Border Terrier, Lhasa Apso or Pug look-alikes.

Conclusion

Chiari-like malformation and syringomyelia occurs in toy breed look-alikes. In Cavalier King Charles spaniel look-alikes the CM prevalence found (96%) resembles the percentage found in previous studies on pedigree Cavalier King Charles spaniels (95%). There has not been enough data concerning the other look-alike toy breeds to draw any conclusions. We can only conclude CM as well as SM does appear in Chihuahua, French Bulldog, King Charles Spaniel, Maltese dog and Jack Russel Terrier look-alikes.

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