Chiari-like malformation, in which the cerebellum is caudally displaced into the foramen magnum, often with concomitant syringomyelia, has been widely diagnosed in CKCSs since it was first recognized in the late 1990s. 1 Nevertheless, the pathogenesis of syringomyelia in CKCSs with Chiari-like malformation is not well understood. The high prevalence of Chiari-like malformation in CKCSs may suggest that affected dogs have a genetic defect similar to the defect that causes type 1 Chiari malformation in humans, 2 which results in underdevelopment of the occipital bone and posterior fossa with subsequent herniation of the caudal cerebellar vermis and obstruction of the foramen magnum. Such abnormalities are thought to interfere with movement of CSF through the caudal fossa, leading to secondary syringomyelia. 3-5 Various fluid dynamic theories have been suggested as mechanisms for the subsequent development of intra-axial cavitation and syringomyelia. 4 However, the specific reasons why syringomyelia might develop in CKCSs are not known, although stenosis or abnormal widening of the foramen magnum, vertebral canal, or both may play a role. 6 A previous study 7 in which a large number of CKCSs underwent magnetic resonance imaging revealed that most had Chiari-like malformation, but that not all affected animals had syringomyelia. Thus, additional studies are needed to determine why some CKCSs with Chiari-like malformation develop syringomyelia and others do not.

Anecdotally, we have previously noticed on magnetic resonance images of some CKCSs a ventrally bulging, hypointense structure located dorsal to the spinal cord between the atlas and the axis, which we thought might represent a hypertrophied dorsal atlantoaxial ligament. In addition, results of radiographic examination suggested that some CKCSs may have subclinical luxation of the atlantoaxial joint, evident as excessive widening of the atlantoaxial joint space on lateral radiographic projections of the cervical region taken with the neck flexed. If dorsal atlantoaxial ligament hypertrophy were to develop as a consequence of mild atlantoaxial joint subluxation and if the hypertrophied ligament were to...
adversely affect CSF movement, then atlantoaxial joint subluxation may represent a possible mechanism for the development of syringomyelia in this breed.

In the present study, therefore, we wanted to test whether excessive widening of the atlantoaxial joint space might be associated with development of syringomyelia in CKCSs. Specifically, the purposes of the study reported here were to determine whether there are significant differences in radiographic morphology of the atlantoaxial region between CKCSs and dogs of other breeds and whether there is an association between radiographic morphology of the atlantoaxial region and syringomyelia in CKCSs.

**Materials and Methods**

Sixty-five CKCSs > 1 year old that underwent magnetic resonance imaging of the brain and cervical portion of the spinal cord at the Queen's Veterinary School Hospital or Stone Lion Veterinary Hospital were included in the study. In addition to magnetic resonance imaging, all dogs had undergone radiography of the cervical region, with lateral radiographic projections obtained with the neck in a neutral position (ie, the position the neck would be in when the dog was walking normally) and a flexed position.

For comparison purposes, radiographs of 72 dogs of other breeds were also examined. Forty-eight of these dogs weighed < 15 kg and were classified as small-breed dogs. This included 7 Jack Russell Terriers, 6 Yorkshire Terriers, 5 Beagles, 4 Dachshunds, 3 Cocker Spaniels, 3 West Highland White Terriers, 2 mixed-breed dogs, and 18 other dogs representing 16 other breeds. The 24 dogs that weighed ≥ 15 kg represented 17 breeds, in addition to 2 mixed-breed dogs. For all 72 dogs, lateral radiographic projections of the cervical region obtained with the neck in a neutral position were available for review. In addition, for 16 of the small-breed dogs, lateral radiographic projections of the cervical region obtained with the neck in a flexed position were also available for review.

**Measurements of radiographic morphology**—For all dogs, the distance that the spinous process of the axis overlapped the dorsal arch of the atlas was measured on the lateral radiographic projection of the cervical region obtained with the neck in a neutral position. This overlap was calculated by measuring the distance between 2 lines drawn perpendicular to the dorsal border of the vertebral foramen of the axis. The first line was drawn at the caudal border of the dorsal arch of the atlas; the second was drawn at the most cranial border of the spinous process of the axis (Figure 1). Overlap was assigned a negative value.

To determine whether any differences in overlap between the atlas and the axis among groups of dog could have resulted from differences in radiographic positioning, the angle between the head and neck was measured on all radiographs. This angle was defined as the angle between a line drawn parallel to the base of the skull and a line drawn parallel to the dorsal border of the vertebral foramen of the axis (Figure 1).

To determine whether any difference in overlap between the atlas and the axis in CKCSs and dogs of other breeds could have been a result of differences in relative size of the spinous process of the axis, relative size of the spinous process of the axis was calculated for all radiographs. Relative size of the spinous process of the axis was calculated by dividing the length of the spinous process of the axis, measured along a line joining the most cranial point of the process with the most caudal point, with the length of the dorsal lamina of the axis (Figure 1).

To determine whether atlantoaxial joint subluxation was more common in CKCSs than in dogs of other breeds, the amount of widening of the atlantoaxial joint that occurred when the neck was moved from a neutral to a flexed position was calculated for those dogs for which both radiographic projections were available for review. For this calculation, the distance between the caudal border of the dorsal arch of the atlas and the cranial border of the dorsal lamina of the axis was measured on both lateral radiographic projections (Figure 1), and the difference was obtained.

**Statistical analysis**—The distribution within compared data sets was tested for normality with the Kolmogorov-Smirnov test. Student t tests were used to compare the following in normal populations: overlap of the atlas and axis, relative size of the axial spinous process, and radiographic angle between the skull and axis between CKCSs and dogs of other breeds. Variances between populations were compared with an F test. The Mann-Whitney test was used for comparisons of populations with unequal variances, such as atlantoaxial joint space widening during flexion. The χ² test was used to determine, for CKCSs, whether excessive atlantoaxial joint space widening (present vs absent) or an excessively small axial spinous process (present vs absent) was significantly associated with syringomyelia (present vs absent). Standard software was used for all analyses.* Values of P < 0.05 were considered significant.

**Results**

**Radiographic morphology**—Overlap between the atlas and axis was significantly (P < 0.001) smaller in CKCSs and dogs of other breeds compared with that in dogs of other breeds. Differences in overlap were significant between the atlas and axis in CKCSs and dogs of other breeds (P < 0.001). This difference was more evident in CKCSs than in dogs of other breeds (P < 0.001).

**Association of radiographic morphology and syringomyelia**—On the basis of magnetic resonance imaging findings, CKCSs were classified as having or not having syringomyelia. Cavalier King Charles Spaniels in which the amount of widening of the atlantoaxial joint that occurred when the neck was moved from a neutral to a flexed position exceeded the upper 95% confidence limit for dogs of other breeds were classified as having excessive atlantoaxial joint space widening. Likewise, CKCSs in which relative size of the spinous process of the axis was less than the lower 95% confidence limit for dogs of other breeds were classified as having an excessively small axial spinous process.

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the 65 CKCSs (mean ± SD, 0.21 ± 0.16 cm) than in all 72 dogs of other breeds (0.71 ± 0.40 cm). In addition, overlap between the atlas and axis was significantly (P < 0.001) smaller in the 65 CKCSs than in the 48 small-breed dogs of other breeds (0.56 ± 0.35 cm). The latter finding did not appear to be a result of differences in radiographic positioning because the angle between the head and neck for the CKCSs was not significantly (P = 0.14) different from the angle for the small-breed dogs of other breeds.

Relative size of the spinous process of the axis was significantly (P < 0.001) smaller in the 65 CKCSs (2.11 ± 0.22 cm) than in all 72 dogs of other breeds (2.49 ± 0.41 cm). Similarly, relative size of the spinous process of the axis was significantly (P < 0.001) smaller in the 65 CKCSs than in the 48 small-breed dogs of other breeds (2.42 ± 0.41 cm).

The amount of widening of the atlantoaxial joint that occurred when the neck was moved from a neutral to a flexed position was not significantly (P = 0.38) different between CKCSs (0.19 ± 0.12 cm; n = 65) and small-breed dogs of other breeds (0.18 ± 0.18 cm; 16). However, the variance was significantly (P = 0.02) different between populations, in that 2 of the small-breed dogs of other breeds (a Cocker Spaniel and a Dachshund) had a large amount of widening (0.5 and 0.6 cm, respectively) and 9 of the 16 (56%) had widening of ≤ 0.1 cm (Figure 2). In contrast, 25 of the 65 (38%) CKCSs had widening of ≤ 0.1 cm, and 19 (29%) had widening of ≥ 0.3 cm.

Differences in the extent of widening of the atlantoaxial joint between populations did not appear to be a result of differences in radiographic positioning, in that the degree of neck flexion (ie, the difference in angle between the head and neck on radiographic views obtained in the neutral and flexed position) and the angle between the head and neck on radiographs obtained in the flexed position were not significantly different between groups (P = 0.82 and 0.48, respectively).

Association of radiographic morphology and syringomyelia—On examination of magnetic resonance images, 60 of the 65 CKCSs could be definitively allocated a syringomyelia status (5 were equivocal and therefore eliminated from analysis). Of these, 21 of the 60 (35%) CKCSs had syringomyelia and 39 (65%) did not. For dogs of other breeds, the upper 95% confidence limit for the amount of widening of the atlantoaxial joint that occurred when the neck was moved from a neutral to a flexed position was 0.3 cm, and on the basis of this cutoff, 6 of the 60 (10%) CKCSs were considered to have excessive atlantoaxial joint space widening (ie, > 0.3 cm). Overall, 3 of the 21 CKCSs with syringomyelia had excessive atlantoaxial joint space widening and 18
did not. In addition, 3 of the 39 CKCSs without syringomyelia had excessive atlantoaxial joint space widening and 36 did not. There was no significant (P = 0.72) association between syringomyelia and excessive atlantoaxial joint space widening.

Similarly, for dogs of other breeds, the lower 95% confidence limit for relative size of the spinous process of the axis was 2.3 cm, and on the basis of this cutoff, 44 of the 60 (73%) CKCSs were considered to have an excessively small axial spinous process (ie, ≤ 2.3 cm). Overall, 17 of the 21 CKCSs with syringomyelia had an excessively small axial spinous process and 4 did not. In addition, 27 of the 39 CKCSs without syringomyelia had an excessively small axial spinous process and 12 did not. There was no significant (P = 0.62) association between syringomyelia and an excessively small axial spinous process.

Discussion

Results of the present study suggested that radiographic morphology of the atlantoaxial region in CKCSs was significantly different from morphology of that region in dogs of other breeds. Specifically, the amount of overlap of the atlas and axis and the relative size of the spinous process of the axis were significantly smaller in CKCSs than in dogs of other breeds. On the other hand, the amount of widening of the atlantoaxial joint that occurred when the neck was moved from a neutral to a flexed position was not significantly different between CKCSs and dogs of other breeds, and contrary to our hypothesis, there was no association between syringomyelia and excessiveatlantoaxial joint space widening or between syringomyelia and an excessively small axial spinous process. We believe that differences in radiographic positioning between groups did not affect these findings because we did not find any significant differences between groups in regard to the angle of the head and neck.

It has previously been stated that consideration of the atlantoaxial articulation would be incomplete if the atlanto-occipital articulation were not considered simultaneously because the 2 joint cavities communicate. Given the high proportion of CKCSs with malformation of the occipital bone, it was not surprising, therefore, that radiographic morphology of the atlantoaxial region differed between CKCSs and dogs of other breeds. Developmental abnormalities of the craniovertebral region are uncommon in dogs, yet multiple malformations are clearly evident in CKCSs. In fact, most CKCSs have Chiari-like malformation, albeit ranging in severity, and in the present study, we found that most also have a relatively small spinous process of the axis. Although excessive atlantoaxial joint widening and an excessively small spinous process of the axis were not associated with syringomyelia in the present study, the presence of so many malformations in the cervical vertebral column suggests the possibility that susceptibility to syringomyelia could be a co-related developmental abnormality. Many variations of skull shape are apparent in various breeds of dog, and some brachycephalic dogs are known to have a prominent dorsal notch in the foramen magnum. It is thought that the further a breed digresses from the ancestral wolf type, the more likely distortions are to be found.

The high proportion of small-breed dogs in the present study with widening of the atlantoaxial joint space when the neck was moved from a neutral to a flexed position was surprising. Atlantoaxial joint subluxation is often diagnosed simply on the basis of excessive distance between the dorsal arch of the atlas and the spinous process of the axis or between the dorsal arch of the atlas and the dorsal limit of the vertebral foramen of the axis. To our knowledge, there are no previous reports on the extent of movement between the atlas and axis that can be expected in healthy dogs. Anatomically, the atlantoaxial joint should be capable of only complex rotational movement with no appreciable flexion, but in the present study, an appreciable degree of atlantoaxial joint space widening was found in many apparently normal individuals. In humans, the degree of mobility of the atlantoaxial joint is determined by measuring the atlantodens distance. This would be a more appropriate measurement in dogs suspected to have atlantoaxial joint instability because it is the rotation of the dens dorsally from the body of the atlas that causes compression of the spinal cord and subsequent clinical signs.

Development of the axial skeleton and development of the CNS are known to be mutually dependent in dogs. Malformations of the axial skeleton and spinal cord have been identified in CKCSs, and although syringomyelia did not appear to be directly associated with the radiographic abnormalities identified in the present study, the coexistence of Chiari-like malformation and syringomyelia suggests widespread maldevelopment of the cranial cervical region. Further work is required to determine why only some CKCSs develop syringomyelia despite the high prevalence of cranial and cervical maldevelopment in the breed.

References

1. Rusbridge C. Persistent scratching in Cavalier King Charles Spaniels. Vet Rec 1997;141:179.


