A PROPOSED RADIOGRAPHIC CLASSIFICATION SCHEME FOR CONGENITAL THORACIC VERTEBRAL MALFORMATIONS IN BRACHYCEPHALIC "SCREW-TAILED" DOG BREEDS

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Congenital vertebral malformations are common in brachycephalic "screw-tailed" dog breeds such as French bulldogs, English bulldogs, Boston terriers, and pugs. The aim of this retrospective study was to determine whether a radiographic classification scheme developed for use in humans would be feasible for use in these dog breeds. Inclusion criteria were hospital admission between September 2009 and April 2013, neurologic examination findings available, diagnostic quality lateral and ventro-dorsal digital radiographs of the thoracic vertebral column, and at least one congenital vertebral malformation. Radiographs were retrieved and interpreted by two observers who were unaware of neurologic status. Vertebral malformations were classified based on a classification scheme modified from a previous human study and a consensus of both observers. Twenty-eight dogs met inclusion criteria (12 with neurologic deficits, 16 with no neurologic deficits). Congenital vertebral malformations affected 85/362 (23.5%) of thoracic vertebrae. Vertebral body formation defects were the most common (butterfly vertebrae 6.6%, ventral wedge-shaped vertebrae 5.5%, dorsal hemivertebrae 0.8%, and dorso-lateral hemivertebrae 0.5%). No lateral hemivertebrae or lateral wedge-shaped vertebrae were identified. The T7 vertebra was the most commonly affected (11/28 dogs), followed by T8 (8/28 dogs) and T12 (8/28 dogs). The number and type of vertebral malformations differed between groups ($P = 0.01$). Based on MRI, dorsal, and dorso-lateral hemivertebrae were the cause of spinal cord compression in 5/12 (41.6%) of dogs with neurologic deficits. Findings indicated that a modified human radiographic classification system of vertebral malformations is feasible for use in future studies of brachycephalic "screw-tailed" dogs.

Key words: butterfly vertebra, congenital vertebral malformation, hemivertebra, kyphosis, scoliosis.

Introduction

HEMIVERTEBRAE, WEDGE-SHAPED (or cuneiform) vertebrae, and butterfly vertebrae are relatively common congenital vertebral malformations in dogs, and are associated with varying degrees of kyphosis and scoliosis.[1,2] Although any breed can be affected, they appear to be more common in brachycephalic "screw-tailed" breeds such as the English bulldog, French bulldog, Boston terrier, and Pug.[1–5] It has been suggested that the kinked tail in these breeds is due to hemivertebra affecting the coccygeal region and this trait has been selected as a desirable phenotype for many generations.[3,4,6] These congenital vertebral malformations can occur in isolation or be multiple in an individual dog, and they occur more frequently in the thoracic vertebral column.[2–5] In general, these malformations are considered to be an incidental radiographic finding because many affected dogs do not show any neurological signs.[1,2,4,5] When clinical signs do occur they are considered to be a consequence of progressive vertebral canal stenosis and vertebral column instability. Vertebral lesions may be apparent at birth, but clinical signs mostly only develop later in life.[1,2,7] The etiology of congenital vertebral malformations remains unclear, but may be due to congenital absence of vertebral vascularization, genetic defects or teratogenic insult to active cartilaginous proliferation.[1,2,4]

In humans, congenital vertebral malformations have been classified into categories on the basis of their radiographic appearance.[9–11] The first classification category includes failure of vertebral segmentation, in which portions

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TABLE 1. Clinical 0 to 5 Points Grading Scale for Thoracolumbar Spinal Cord Lesions[13]

Grade	Clinical signs
0	Normal
1	Spinal pain
2	Ambulatory paraparesis
3	Nonambulatory paraparesis
4	Paraplegia with intact deep pain perception
5	Paraplegia with absent deep pain perception

of adjacent vertebral elements fail to divide (block vertebra or bars). The second category consists of failure of vertebral formation, in which a portion of a vertebral element is deficient (hemivertebra, wedge shape vertebra or butterfly vertebra). Based on these categories, Nasca et al.[10] proposed a radiographic classification for congenital scoliosis and McMaster et al.[11] for congenital kyphosis and kyphoscoliosis. One previous canine study used the Nasca classification system in five clinical cases and one canine abstract classified wedging of the vertebral body as unilateral, dorsal, and ventral.[5,12] To the authors' knowledge, no study has evaluated the McMaster radiographic classification scheme for use in brachycephalic "screw-tailed" dogs.

In this retrospective study, our primary objectives were (1) to develop and apply a radiographic classification scheme for congenital vertebral malformations affecting the thoracic vertebral column in a group of brachycephalic "screw-tailed" dogs and (2) to test associations between the type of vertebral malformation and the presence of neurological deficits.

Materials and Methods

The medical records of the University of Glasgow Small Animal Hospital were retrospectively reviewed from September 2009 to April 2013 to identify French bulldogs, English bulldogs, Boston terriers, and Pugs with or without neurological deficits, that had well positioned lateral and ventro-dorsal digital radiographs of the thoracic vertebral column, with at least a single vertebral congenital malformation present. The age, sex, and breed were recorded. If any neurological deficits were associated with the vertebral malformation, then the duration of clinical signs and the neurological grade at presentation were recorded. The neurological grade was determined using the standard 0 to 5 grading system (Table 1)[13]. Patients were then divided into two groups, one with neurological deficits localized to the thoracic spinal cord segments (Group 1) and one without neurological deficits (Group 2).

Radiographs for each dog were retrieved and evaluated by two observers who were unaware of dog group status at the time of interpretation. Images were displayed using an open-source PACS Workstation DICOM viewer (Osirix Imaging Software, v 3.9.2, Pixmeo, Geneva, Switzerland). Each vertebral malformation classification was determined by a consensus between the two observers. The number and the vertebral level were also recorded for each vertebral malformation and for each patient. When available, MRI studies were also retrieved and interpreted.

The vertebral malformations were classified as defects of segmentation if adjacent vertebral elements fail to divide (block vertebrae) or defects of formation if a portion of the vertebra was deficient. Defects of formation were then subclassified into ventral aplasia of the vertebral body (dorsal hemivertebra), lateral aplasia of the vertebral body (lateral hemivertebra), ventro-lateral aplasia of the vertebral body (dorso-lateral hemivertebra), ventral and median aplasia of the vertebral body (butterfly vertebra), ventral hypoplasia of the vertebral body (ventral wedge shape vertebra), and lateral hypoplasia of the vertebral body (lateral wedge shape vertebra) using a modified classification based on that of McMaster et al.[11] (Fig. 1). The presence of any other vertebral malformations, including thoracic and cervical transitional vertebrae, spina bifida and any other types, were also recorded.

Statistical tests were selected by a statistician. Data were analyzed using commercially available software (Minitab 16 Statistical Software, Minitab Inc. State College, PA, USA). Descriptive statistics were reported as mean, range, and standard deviation (SD). A Fisher exact test was used to compare the vertebral malformations between the two groups with significance defined as $P < 0.05$. For that the number of dogs with severe vertebral malformations (ventral aplasia of the vertebral body, lateral aplasia of the vertebral body, and ventro-lateral aplasia of the vertebral body) and with less severe vertebral malformations (ventral and median aplasia of the vertebral body, ventral hypoplasia of the vertebral body, lateral hypoplasia of the vertebral body, and other unclassified vertebral malformations) in each of the groups was calculated.

Results

Twenty-eight dogs were included in the study. Eleven Pugs, 11 English bulldogs, three French bulldogs, and three Boston terriers. The mean age was 2.5 years (range: 4 months - 14 years, SD: 2.86 years). There were 20 males (two neutered) and eight females (one neutered). Twelve dogs had neurological deficits (Group 1) and 16 dogs did not have neurological deficits (Group 2) at the time of thoracic spinal radiography. The 12 dogs with neurological deficits (Group 1) included nine Pugs, two English bulldogs and one French bulldog. The mean age was 2.4 years (range: 4 months - 7.5 years, SD: 2.4 years). There were six males (one neutered) and six females (one neutered). Nine dogs presented with ambulatory paraparesis and ataxia

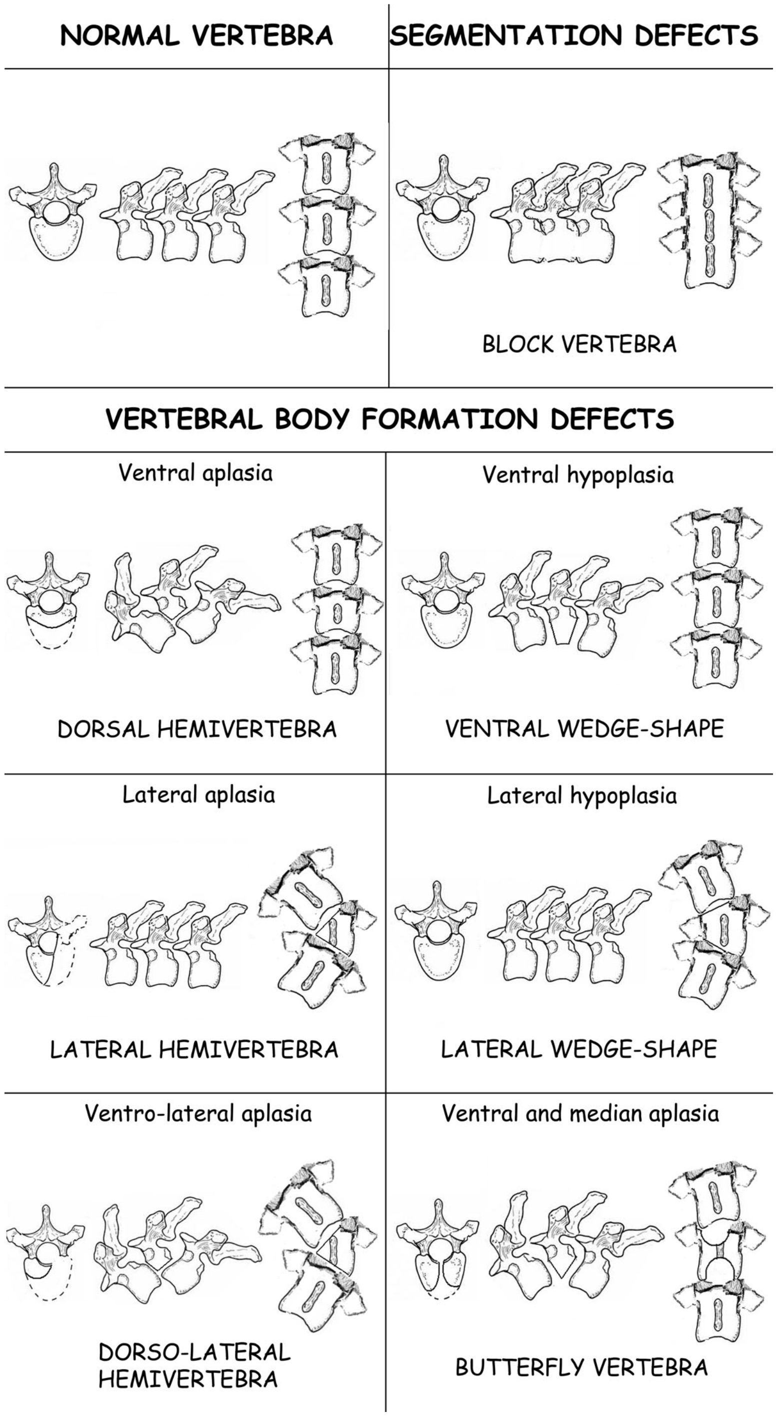

FIG. 1. Schematic line drawing showing the radiographic appearance of the vertebral malformation classification system used in this study.

(grade 2), two with nonambulatory paraparesis (grade 3) and one with paraplegia (grade 4). The mean duration of neurological deficits prior to presentation was 68 days (range: 1–240 days, SD: 69 days). All Group 1 dogs had MRI of the thoracic spine using the same 1.5 Tesla scanner (Magnetom Essenza, Siemens, Camberley, United Kingdom), and spinal cord compression was identified in all of them. Sagittal T2-weighted images and transverse T2-weighted and T1-weighted images of the area of interest were acquired in all these cases. In 10 of the dogs, MRI confirmed that spinal cord compression was directly caused by the vertebral malformation with no other

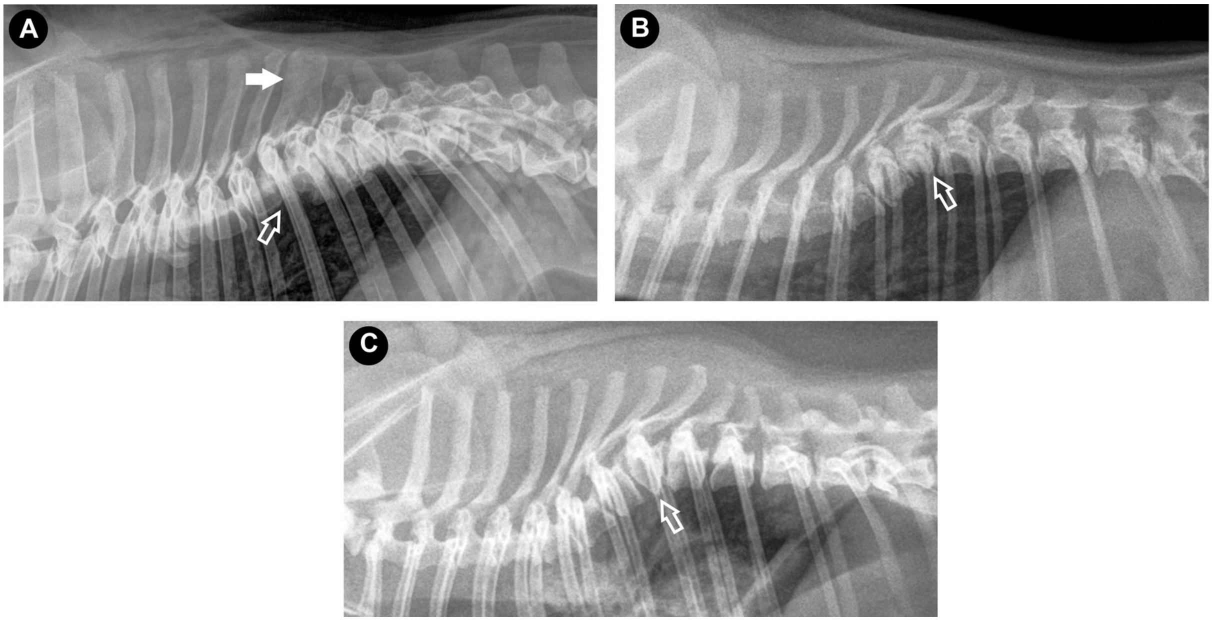

FIG. 2. Congenital vertebral malformations identified on lateral radiographs of the thoracic vertebral column. (A) Block vertebrae, where there is failure of normal segmentation of the vertebrae, evident as the failure of division of T8 and T9 vertebral bodies (open arrow) and spinous process (solid arrow). (B) Ventral hypoplasia of T8 vertebral body (ventral wedge shape vertebra) (arrow) with kyphosis. (C) Ventral aplasia of T8 vertebral body (dorsal hemivertebra) (arrow) with more severe kyphosis.

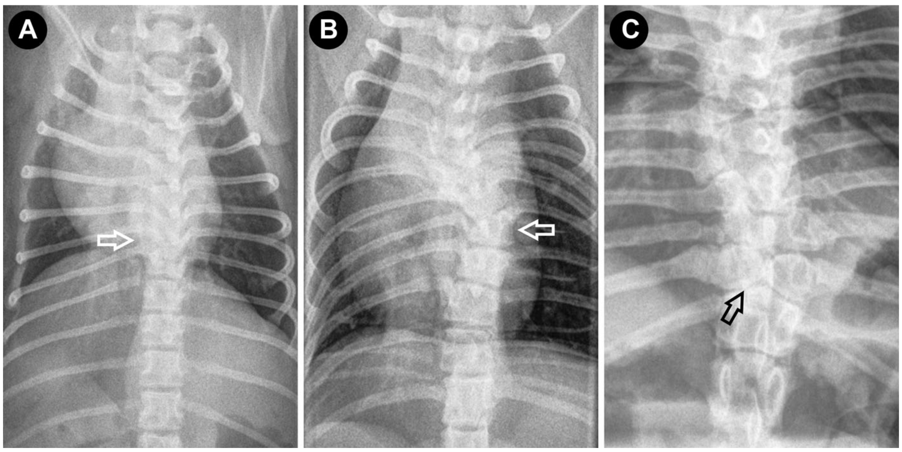

FIG. 3. Congenital vertebral malformations identified on ventro-dorsal radiographs of the thoracic vertebral column. (A) Ventral hypoplasia of T8 vertebral body (arrow, this is the same case as figure 2B): no evidence of scoliosis is evident on the ventro-dorsal view if there is no lateral hypoplasia. (B) Ventro-lateral aplasia of T8 vertebral body (dorso-lateral hemivertebra) (arrow). This case had similar changes to figure 2C on the lateral view. (C) Ventral and median aplasia of the vertebral body (Butterfly vertebra) (arrow).

findings. One dog had an intervertebral disc herniation adjacent to the vertebral malformation and another dog had discospondylitis and secondary empyema in the intervertebral disc adjacent to the vertebral malformation. The 16 dogs with no neurological deficits (Group 2) included nine English bulldogs, three Boston terriers, two Pugs, and two French bulldogs. The mean age was 2.6 years (range: 7 months - 14 years, SD: 3.2 years). There were 14 males (one neutered) and two females. None of the Group 2 dogs had MRI of the thoracic spine.

In all dogs, radiography of the thoracic vertebral column was performed using the same digital radiography system (Siemens, Camberley, United Kingdom). A total of 362 vertebrae were evaluated, as two of the dogs had 12 thoracic vertebrae. A congenital malformation was identified in 85 (23.5%) vertebrae, with 18 (64.2%) of the dogs having multiple malformations. It was possible to classify 50 of these vertebral malformations using the proposed classification (Fig. 1). Of the 35 vertebral malformations that failed to meet the criteria for classification, 26 vertebrae

TABLE 2. Number of Vertebrae Affected by Each Type of Congenital Vertebral Malformation in Dogs with Associated Neurological Deficits (Group 1) and Dogs Without Associated Neurological Deficits (Group 2). A Significant Difference in the Types of Congenital Vertebral Malformations was Evident when Comparing the Two Groups ($P = 0.01$)

Vertebral malformation	All dogs	Group 1	Group 2
Block vertebra	1	1	0
Ventral aplasia of the vertebral body (Dorsal hemivertebra)	3	3	0
Ventro-lateral aplasia of the vertebral body (Dorso-lateral hemivertebra)	2	2	0
Lateral aplasia of the vertebral body (Lateral hemivertebra)	0	0	0
Ventral and median aplasia of the vertebral body (Butterfly vertebra)	24	4	20
Ventral hypoplasia of the vertebral body (Ventral wedge shape)	20	6	14
Lateral hypoplasia of the vertebral body (Lateral wedge shape)	0	0	0
Total number of vertebrae with a classified congenital malformation	50	16	34
Total number of vertebrae with an unclassified congenital malformation	35	14	21
Total number of vertebrae evaluated	362	154	208

were judged as having a shorter vertebral body length with a normal shape, four had fused spinous processes with normal vertebral bodies, three were transitional vertebrae and two demonstrated spina bifida. Of the 50 vertebral malformations classified, one was a defect of vertebral segmentation and 49 were defects of vertebral formation. The most common malformation was butterfly vertebra seen in 24 vertebrae (6.6%), followed by ventral wedge shape vertebra in 20 vertebrae (5.5%), dorsal hemivertebra in three vertebrae (0.8%), dorso-lateral hemivertebra in two vertebrae (0.5%) and block vertebra in one vertebra (0.3%) (Figs. 2 and 3). No lateral hemivertebra or lateral wedge shape vertebra were identified in this study (Table 2). The most commonly malformed vertebra was T7 (11 dogs), followed by T8 (eight dogs) and T12 (eight dogs). Figure 4 demonstrates the distribution of the type of malformation by vertebral column level.

There was a significant difference in the number of vertebral malformations between groups ($P = 0.01$). Dorsal and dorso-lateral hemivertebrae were only present in Group 1 dogs, and these were interpreted to be the cause of neurological deficits in 5/12 (41.6%) of these dogs. Butterfly and wedge shape vertebrae were identified in both groups, but were more commonly seen in Group 2 dogs with no neurological deficits (Table 2). Butterfly vertebrae were not found to be the cause of the spinal cord compression in MRI studies of any of the 10 dogs where the spinal cord compression was directly caused by the congenital vertebral malformation.

Discussion

In the present study we proposed and applied a radiographic classification of congenital vertebral malformations in dogs based on previous human studies of congenital kyphotic, scoliotic, and kypho-scoliotic malformations.[10,11] This classification scheme was helpful for more consistently describing the different types of vertebral malformations present in these breeds, specifically the defects of vertebral body formation. Similar to humans, defects of vertebral formation were more common in the dogs included in this study than defects of vertebral segmentation.[11] The results of the present study indicated that less severe vertebral body formation defects such as ventral and median aplasia (butterfly vertebra) or ventral hypoplasia of the vertebral body (ventral wedge shape vertebra) are common and can be seen in dogs with and without neurological deficits. Where spinal cord compression was directly caused by the congenital vertebral malformation, none were due to ventral and median aplasia of the vertebral body (butterfly vertebra) in this sample population. More severe defects of vertebral body formation, including ventral and ventro-lateral aplasia of the vertebral body (dorsal and dorso-lateral hemivertebrae), were more likely to be associated with neurological deficits. These findings are similar to human patients, where the severity of defects of vertebral body formation has been shown to be proportional to the severity of the kyphosis.[11] Another interesting finding of the current study was that no lateral aplasia or hypoplasia of the vertebral body was observed. It is possible that kyphosis and kypho-scoliosis are more frequent and important than scoliosis in brachycephalic "screw-tailed" dog breeds. It is also important to notice that of the 35 vertebral malformations that failed to meet the classification criteria, 26 were judged as having a shorter vertebral body length with a normal shape. For future studies, the authors recommend that this type of defect of vertebral body formation should be added to the classification scheme as symmetrical hypoplasia of the vertebral body ("short" vertebra) (Fig. 5).

Findings in the current study were consistent with previous studies indicating that congenital vertebral malformations are common in the thoracic vertebral column of brachycephalic "screw-tailed" dog breeds, with 23.5% of the vertebrae examined in this study being affected. The majority of the dogs (64.2%) in this study had multiple vertebral malformations. This finding was also similar

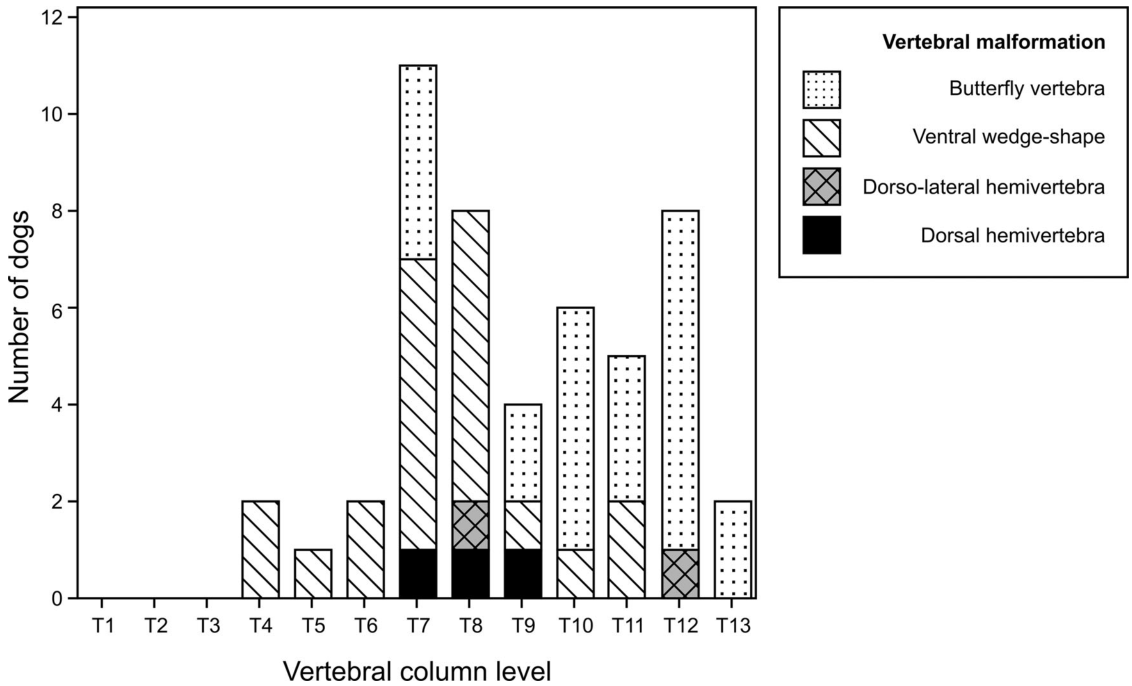

FIG. 4. Histogram showing the frequency of defects of vertebral body formation by vertebral column level.

to previous reports describing a high frequency of vertebral abnormalities in French bulldogs, English bulldogs, and other brachycephalic breeds with multiple vertebrae commonly affected.[2–5] Also similar to previous studies was our observation that the most commonly affected vertebra was in the midthoracic region (T6-T9 region).[1,2,5]

The neurological signs were compatible with a T3-L3 spinal cord segment myelopathy in all the cases with neurological deficits. The majority of dogs with neurological signs (75%) presented with a chronic and progressive history of ambulatory paraparesis and ataxia (Grade 2), which suggested slowly progressive spinal cord injury and/or compression, consistent with previous reports.[1,2,7,14] Pugs were noted to have neurological deficits in 75% of the patient population evaluated here. Two possibilities for the over-representation of neurological deficits in this breed may be that there are more severe vertebral malformations present in this breed, or that they were more common in our population. In two of the 12 cases with neurological deficits, it was found that the clinical signs were not a direct consequence of vertebral canal stenosis caused by the vertebral malformation, but were secondary to another neurological disease that occurred concurrently. In both cases the intervertebral discs adjacent to the vertebral malformation were the ones involved (one dog with Hansen type I intervertebral disc herniation and the other dog with discospondylitis and secondary empyema). It is possible that intervertebral instability between the normal and malformed vertebrae could have resulted in damage to the intervertebral disc and predisposed it to herniation or infection.

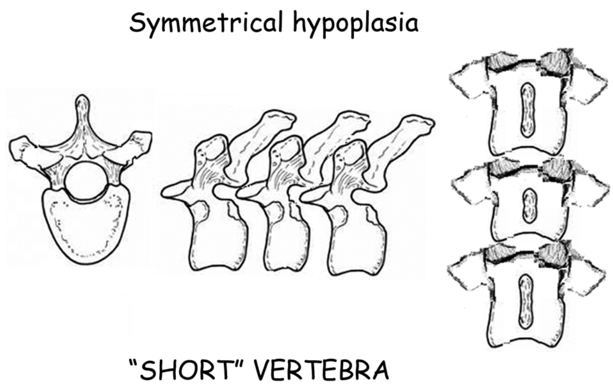

FIG. 5. Schematic line drawing showing the radiographic appearance of the additional vertebral malformation found frequently in this study: symmetrical hypoplasia of the vertebral body ("short" vertebra).

There were several limitations to this study, including it being retrospective in nature. Second, the breed distribution between the two groups was not similar and this could have influenced the type of vertebral malformations seen in each group. Third, the number of cases in the present study was small, thus it was not possible to give an accurate representation of the frequency of the different vertebral malformations in each breed. Another limitation was the absence of long-term follow up of the cases and it is unknown if some of the dogs in Group 2 may have developed neurological deficits at a later date. None of them had any additional imaging to confirm that there was no spinal cord compression at the site of their vertebral body anomalies.

The authors feel that this is an important consideration when interpreting the results of this study.

In conclusion, radiographic classification of congenital vertebral malformations in brachycephalic “screw-tailed” dogs is feasible. Defects of vertebral body formation were the most frequent in this sample population. More severe defects of vertebral body formation, such as ventral and ventro-lateral aplasia (dorsal and dorso-lateral hemivertebrae), may more likely be associated with neurological deficits, but further studies in a larger group of dogs are needed to confirm this.

Conflict of Interest Statement

None of the authors has any financial or personal relationships that could inappropriately influence or bias the content of the paper.

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