

FOUR LEG NEWS

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BRACHYCEPHALIC SPINES

If you've been doing canine rehabilitation / physiotherapy for any period of time, you're bound to have run into a wobbly Frenchie or foot-dragging pug. This newsletter delves into the brachycephalic spine and the plethora of abnormalities that might be found there! I found this topic fascinating, so I'm sure that my fellow nerds will find it just as enthralling as I did! Enjoy!

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**GUTIERREZ-QUINTANA, R. AND DE DECKER, S.
(2021), TAIL END OF THE BRACHYCEPHALIC
PROBLEM: DIAGNOSTIC AND TREATMENT OPTIONS
FOR SPINAL MALFORMATIONS. IN PRACTICE, 43:
124-134.**



"Vertebral malformations are very common in small brachycephalic breeds, such as French bulldogs, English bulldogs, Boston terriers and pugs. In many cases they are considered incidental findings, but in others they can be associated with progressive neurological deficits."

"As they are common incidental radiographic findings, it can be very difficult and challenging to identify the clinically relevant ones."

Despite a similar 'look', pugs do not have a "screw-tail" and belong to a completely different genetic clade and as such have different types, frequencies, and location of vertebral malformations.

BOX 1: VERTEBRAL AND SPINAL MALFORMATIONS IN SMALL BRACHYCEPHALIC BREEDS

Spina bifida

- Pugs: Common incidental finding in cranial thoracic region
- French and English bulldogs: Rare, but when present at L7–S1 it can be associated with meningocele, meningomyelocele and neurological deficits

Vertebral body formation defects (hemivertebrae, butterfly vertebrae)

- Pugs: Rare, but when present likely to be clinically significant in young dogs
- French and English bulldogs: Common incidental finding, rarely causing neurological deficits
- Severe kyphosis is more likely to be clinically significant

Caudal articular process aplasia/hypoplasia

- Pugs: Common finding affecting caudal thoracic and thoracolumbar region. It has been associated with spinal arachnoid diverticula, meningeal fibrosis and chronic intervertebral disc protrusions causing neurological deficits
- French and English bulldogs: Common incidental finding

Transitional vertebra

- Pugs: Common incidental finding in cervicothoracic, thoracolumbar and lumbosacral region
- French and English bulldogs: Uncommon incidental finding in lumbosacral region

Cranial thoracic vertebral canal stenosis

- Pugs: Not reported
- French and English bulldogs: Common incidental finding, but can cause neurological deficits
- Severe stenosis is more likely to be clinically significant

Spinal arachnoid diverticula and meningeal fibrosis

- Subarachnoid diverticula: Pugs and French bulldogs are some of the most commonly affected breeds at the thoracolumbar region and show progressive neurological deficits. Young pugs are affected at the cervical region
- Meningeal fibrosis: Possible breed predisposition in pugs

Descriptions:

Spina bifida is a congenital vertebral malformation in which there is failure of the vertebral arch to close over the spinal cord.

Vertebral body formation

defects refer to malformations of thoracic and lumbar vertebral bodies, secondary to failure of vertebral ossification centre formation during development. Older descriptions for such were termed hemivertebrae or butterfly vertebra, however the more correct description would be aplasia or hypoplasia of the vertebral body.

Cranial thoracic vertebral canal stenosis is a relatively uncommon vertebral malformation in dogs, which has mainly been reported in large and giant breeds.

Spinal arachnoid diverticula

consist of accumulation of cerebrospinal fluid (CSF) within a focal area of the subarachnoid space, suspected to be secondary to the presence of subarachnoid adhesions.

Meningeal fibrosis (also known as constrictive myelopathy) is characterized by the presence of fibrous tissue affecting the dura mater.

Transitional vertebrae can occur at any vertebral column junction and retain characteristics of the two adjacent regions, cervicothoracic, thoracolumbar and lumbosacral region; and they tend to be associated with an abnormal vertebral count.

Caudal articular process hypoplasia and aplasia, affecting the thoracic vertebrae, thus impacting stability of the vertebral column.

TREATMENT:

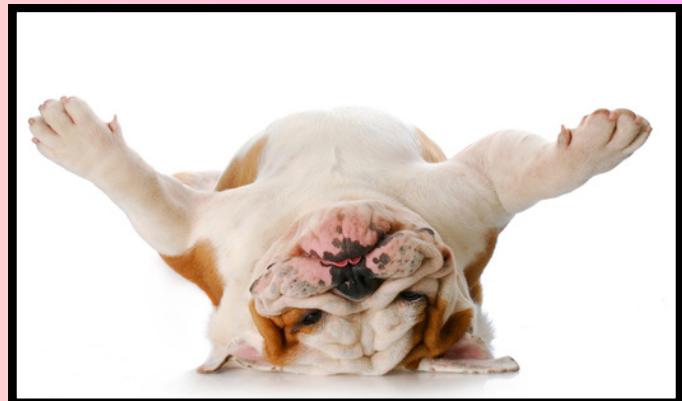
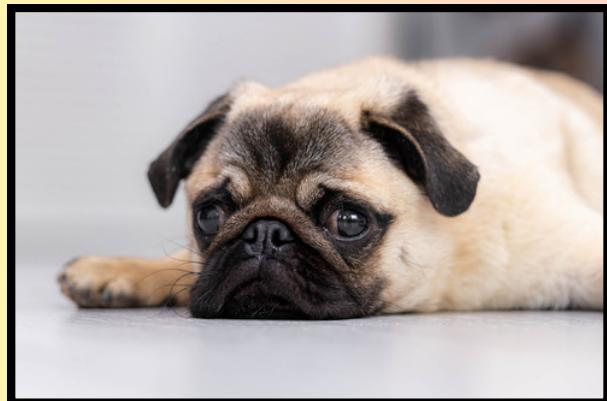
Little information exists regarding the best treatment for spina bifida associated with meningocele or meningomyelocele. A case series of bulldogs with lumbosacral meningocele or meningomyelocele that were treated surgically, the majority of dogs showed no further deterioration of the clinical signs and in some cases even partial improvement.

Vertebral body formation defects causing severe kyphosis are best treated surgically as response to medical management is poor.

The treatment for caudal articular process hypoplasia/aplasia is only necessary if it is associated with other pathologies, such as intervertebral disc disease, arachnoid diverticulum or meningeal fibrosis (also known as constrictive myelopathy).

Cranial thoracic vertebral canal stenosis is usually treated surgically with either a dorsal laminectomy or a hemilaminectomy.

Arachnoid diverticula have been treated medically with anti-inflammatory medications, and surgically using laminectomies with durectomies, durotomies or marsupialisation of the dura. Surgery is generally recommended, as the long-term outcome has been shown to be better.



Mansour TA, Lucot K, Konopelski SE, et al. (2018) Whole genome variant association across 100 dogs identifies a frame shift mutation in DISHEVELLED 2 which contributes to Robinow-like syndrome in bulldogs and related screw tail dog breeds. PLoS Genetics 14, e1007850.

Dogs with a truncated and kinked tail, commonly known as a screw-tail, such as the French bulldog, English bulldog and Boston terrier, are predisposed to vertebral column malformations, and recently a mutation in the Dishevelled 2 gene has been reported as a contributing factor.



De Decker S, Rohdin C, Gutierrez-Quintana R. Vertebral and spinal malformations in small brachycephalic dog breeds: Current knowledge and remaining questions. Vet J. 2024 Mar 6;304:106095.

Understanding the Pathologies

HEMIVERTebra:

Ten (10) hemivertebra subtypes have been recognized. Most common are ventral aplasia or 'dorsal hemivertebra', ventral hypoplasia or 'wedged vertebra', and ventral and median aplasia or 'butterfly vertebra'.

Hemivertebrae are most often found in the mid-thoracic spine. Especially in French and English bulldogs, it is common to find multiple hemivertebrae along the vertebral column. Hemivertebrae can cause kyphosis or scoliosis, and in French bulldogs is associate with the screw-tail morphology.

The prevalence of hemivertebrae in French bulldogs is between 75% and 100%. One study demonstrated one or more thoracic hemivertebrae in 94% of neurologically normal French bulldogs, 73.2% of English bulldogs and 17.6% of pugs. One study reported lumbosacral hemivertebra in 32% of neurologically normal French bulldogs, 24.3% of English bulldogs and 1.7% of pugs.

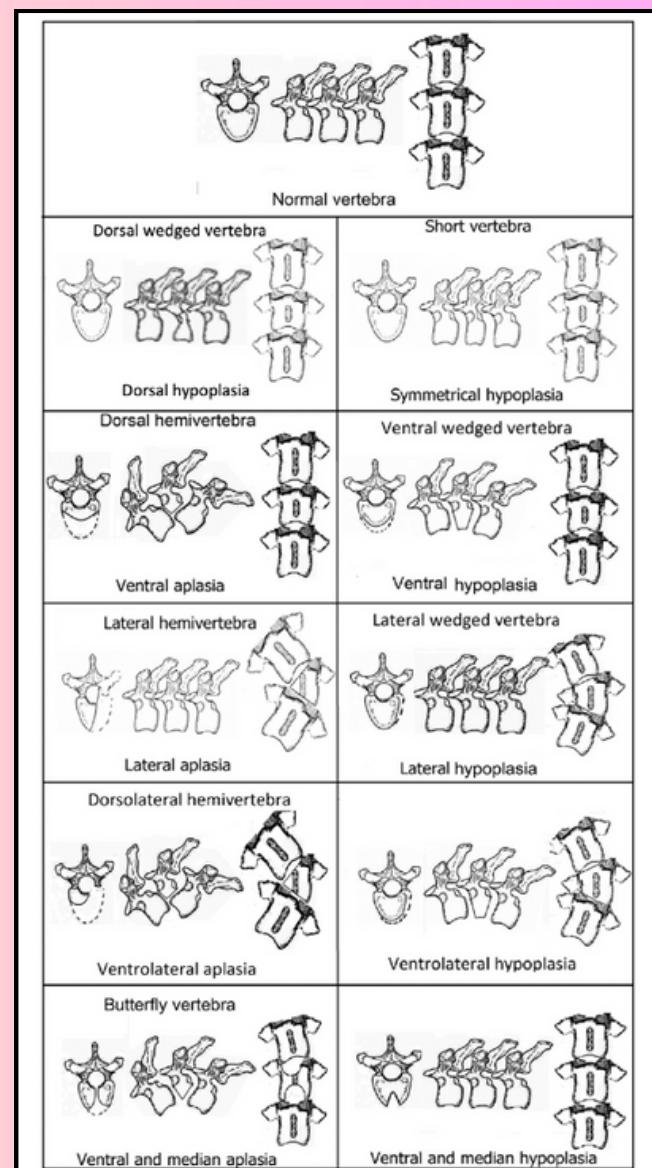


Fig. 1. Diagram of the 10 reported subtypes used to classify canine hemivertebra using computed tomography. This diagram is based on a previously published classification system (Ryan et al., 2019) and expanded to include the subtype dorsal wedged vertebra or dorsal hypoplasia.

Clinical findings

Some dogs with hemivertebra will develop signs of progressive pelvic limb ataxia and paraparesis. Although most affected dogs are younger than one year of age, clinical signs can occur later in life.

In Frenchie's and English bulldogs hemivertebra are considered incidental unless accompanied with kyphosis, as this can predispose to IVDE. French bulldogs with kyphosis were at twice the odds of having a thoracolumbar intervertebral disc extrusion. Studies have suggested that a Cobb angle exceeding 35° is associated with an increased likelihood of clinical signs and measurement of Cobb angles on diagnostic imaging studies is relatively easy and reliable.

Hemivertebra in pugs, do, however tend to be more clinically relevant.

Surgery might help in a case of a hemivertebra causing kyphosis and neurologic signs. Stabilization might have a positive impact.

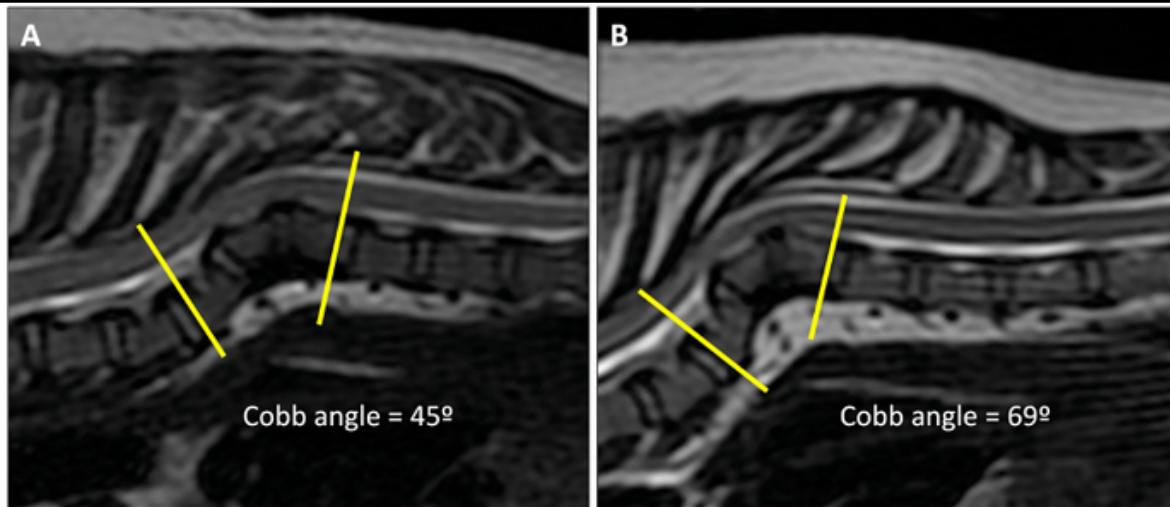


Fig. 2. (A) T2-weighted sagittal MR image of a 5-month-old pug with mild paraparesis and ataxia of the pelvic limbs. A thoracic hemivertebra with kyphosis can be seen. The measured Cobb angle is 45 degrees. (B) The same dog 2 months later. Although still ambulatory, more severe ataxia and paresis is seen. The measured Cobb angle is 69 degrees, indicating worsening of the previously diagnosed kyphosis. Although it is currently unknown how increased kyphosis results in progression of clinical signs, it is possible this is associated with increased instability or a more severe dynamic component of spinal cord compression.

CAUDAL ARTICULAR PROCESS DYSPLASIA

It has been proposed that CAPD may cause low-grade repetitive vertebral instability, which could subsequently lead to reactive fibrotic tissue formation surrounding the spinal cord. The latter has been referred to as constrictive myelopathy or meningeal fibrosis.

Prevalence

There is a high prevalence of CAPD in neurologically normal French bulldogs (70.4%), English bulldogs (81.4%) and pugs (97%). Pugs also exhibit a higher number of affected vertebrae per dog, more often bilateral dysplasia, and CAPD localised in the caudal (vs. cranial) thoracic vertebral segments.

Clinical Relevance:

Studies have been unable to conclusively confirm a causative relationship between CAPD and other myelopathies.

Treatment? Caution should be exercised with surgical interventions as vertebral instability should be resultant with decompressive surgeries.

TRANSITIONAL VERTEBRA

T-L and L-S junctions are most commonly affected. They can result in variations of the number of vertebral segments. Transitional vertebrae are significantly more common in pugs than other small brachycephalic breeds, affecting cervicothoracic, thoracolumbar, and lumbosacral regions.

Pugs are one of the only breeds to have frequent cervical ribs on C7 (46% of neurologically normal pugs). Thoracolumbar and lumbosacral transitional vertebrae are also common in this breed with multiple studies reporting a prevalence higher than 30% for both regions.



THORACIC CANAL STENOSIS

It is observed predominantly in young male English and French bulldogs – most commonly at T2-T3 and T3-T4.

Not all stenosis is clinically relevant. Reported success with both surgery or medical management.

SPINA BIFIDA

The embryonic failure of fusion of one or more vertebral arches; it can happen as a single entity or be accompanied with spinal cord malformations. As a single entity, it is common in pugs at T1, and in French and English bulldogs as well as pugs, it is seen at the L-S junction.

Spina bifida with concurrent meningocele or meningomyelocele in the lumbosacral region has been reported in English and French bulldogs causing urinary and fecal incontinence. Little information exists as to best management.

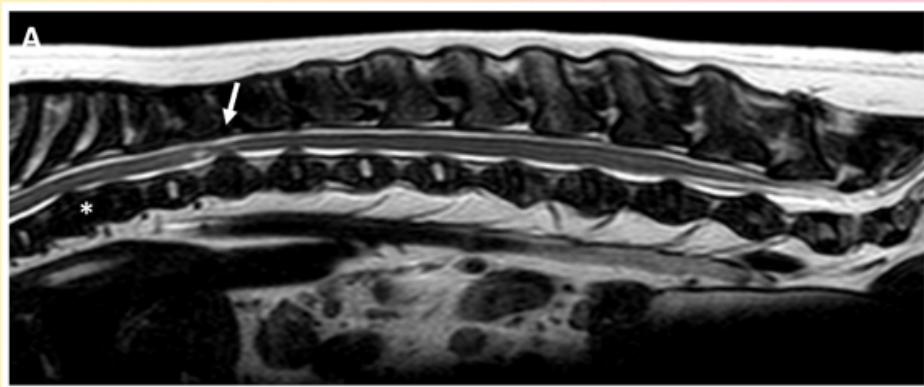
VERTEBRAL VASCULAR CANAL DYSPLASIA

Identified on CT or MRI imaging of French and English bulldogs, is characterized by a defect in the ossification of the vertebral body of variable extent, centred around the vascular canal. Further studies are needed to understand the possible etiology and clinical relevance.

SPINAL MALFORMATIONS

1. Spinal arachnoid diverticula (SAD) – Focal dilations of the subarachnoid space containing cerebrospinal fluid (CSF).
2. Meningeal fibrous adhesion – Described as meningeal fibrosis (MF) or constrictive myelopathy.

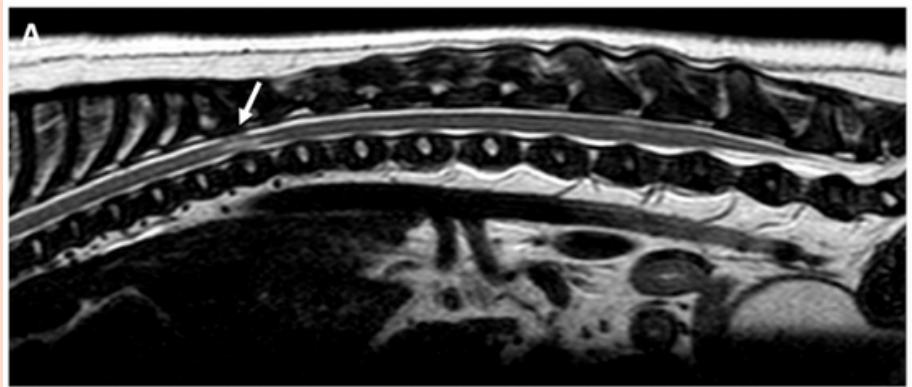
Dogs with SAD and MF present most often with a chronic onset of progressive ataxia and paresis. Urinary or fecal incontinence occurs in approximately 20% of cases and spinal hyperesthesia is typically not present. Especially in pugs, it is common to observe multiple concurrent thoracolumbar spinal conditions, such as SAD, CAPD, MF, and intervertebral disk herniation, when they are assessed for a chronic, progressive T3-L3 myelopathy. In fact, this combination has been referred to 'pug dog thoracolumbar myelopathy'.



(A) T2-weighted sagittal MR image of an 8-year-old pug with chronic progressive paraparesis and ataxia of the pelvic limbs. A vertebral body malformation is present at the level of T9 (asterisk). A mild widening of the dorsal subarachnoid space can be at the level of T11-T12 (arrow). An intraparenchymal hyperintensity and intervertebral disk protrusion can also be observed at this level.

Meningeal fibrosis / constrictive myelopathy is diagnosed by MRI. Meningeal fibrosis is characterized by the formation of a dense, sometimes circumferential, band of fibrotic tissue, causing intradural adhesions between the arachnoid and pia mater and constriction of the spinal cord. Although it can occur in other breeds, MF is heavily associated with the pug breed. Affected animals develop clinical signs later in life, with studies reporting a median age around 7.5 years. It typically affects the caudal thoracic vertebral column.

Treatments: results of medical management are largely unknown, decompressive surgery along with dissolution of the pia-arachnoid adhesions has poor outcomes, stabilization has been associated with neurological improvements, but fecal and urinary incontinence persists in more than half of affected cases.

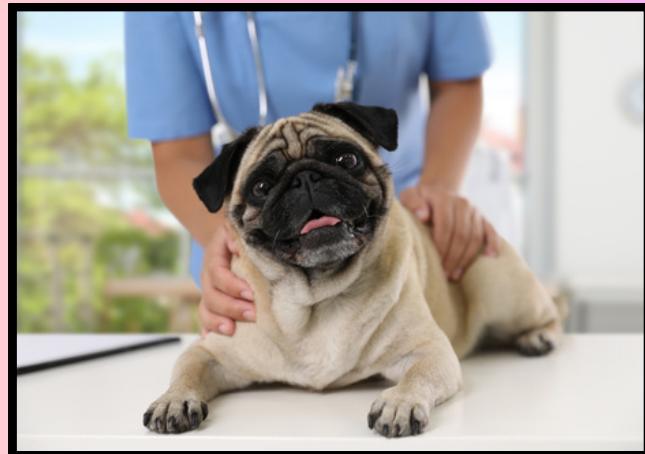


Why do pugs and French bulldogs experience spinal arachnoid diverticula?

Despite their physical similarities, pugs and French bulldogs are not closely related breeds, and as such a genetic component is unlikely. Maternal hypoxia can result in vertebral defects in mammals and hypoxia is also linked to a decreasing ability to maintain normal spinal cord function. Furthermore, an association has been shown between an abnormal gait (ataxia and paresis) and dyspnea in pugs.

Why are pugs affected by meningeal fibrosis?

Pugs are prone to inflammatory CNS disease, such as necrotizing meningoencephalitis (NME). Pugs with focal fibrous adhesions may present with lympho-histiocytic inflammation, similar to dogs with NME. A recent study also recognized a strong inflammatory cytokine expression in a group of older pugs. A dysregulated immune response could potentially interfere with the normal physiological repair process resulting in excessive fibrous tissue.



De Decker S, Packer RMA, Cappello R, et al. Comparison of signalment and computed tomography findings in French Bulldogs, Pugs, and English Bulldogs with and without clinical signs associated with thoracic hemivertebra. J Vet Intern Med. 2019 Sep;33(5):2151-2159.

Pugs and a severe degree of kyphosis should be considered the most important factors when considering a diagnosis of clinically relevant thoracic hemivertebra. Our results further determined that fewer as opposed to more hemivertebrae along the vertebral column more often are associated with clinical signs.

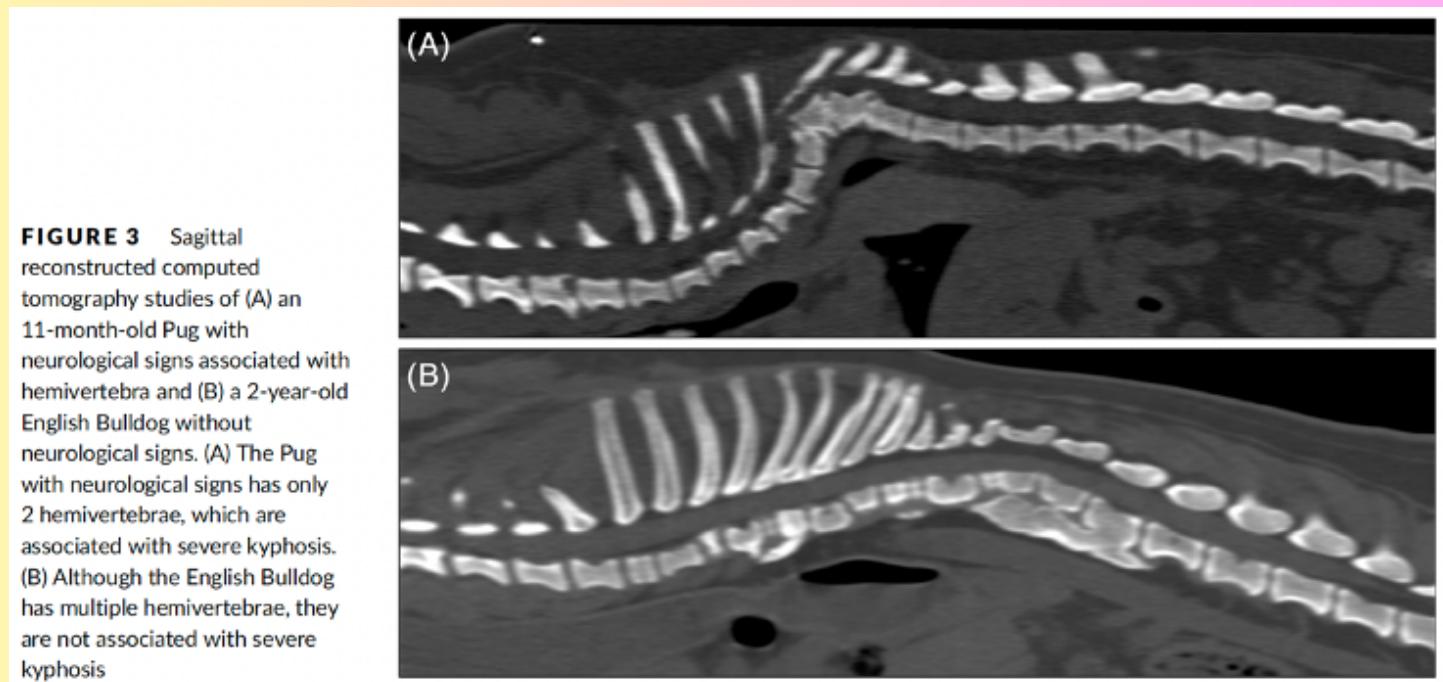


FIGURE 3 Sagittal reconstructed computed tomography studies of (A) an 11-month-old Pug with neurological signs associated with hemivertebra and (B) a 2-year-old English Bulldog without neurological signs. (A) The Pug with neurological signs has only 2 hemivertebrae, which are associated with severe kyphosis. (B) Although the English Bulldog has multiple hemivertebrae, they are not associated with severe kyphosis

What about conservative management?

Outcomes of nonsurgical treatment for congenital thoracic vertebral body malformations in dogs: 13 cases (2009–2016)

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OBJECTIVE

To characterize outcomes following nonsurgical treatment of congenital thoracic vertebral body malformations causing neurologic deficits in dogs.

DESIGN

Retrospective case series.

ANIMALS

13 client-owned dogs treated nonsurgically for congenital thoracic vertebral body malformations at 3 veterinary referral hospitals from June 2009 through May 2016.

PROCEDURES

Data were extracted from the medical records regarding dog signalment, duration and type of clinical signs before referral, general physical and neurologic examination findings, radiographic and MRI findings, and treatments provided after diagnosis. Follow-up data were obtained from records of recheck examinations and via a standardized owner questionnaire.

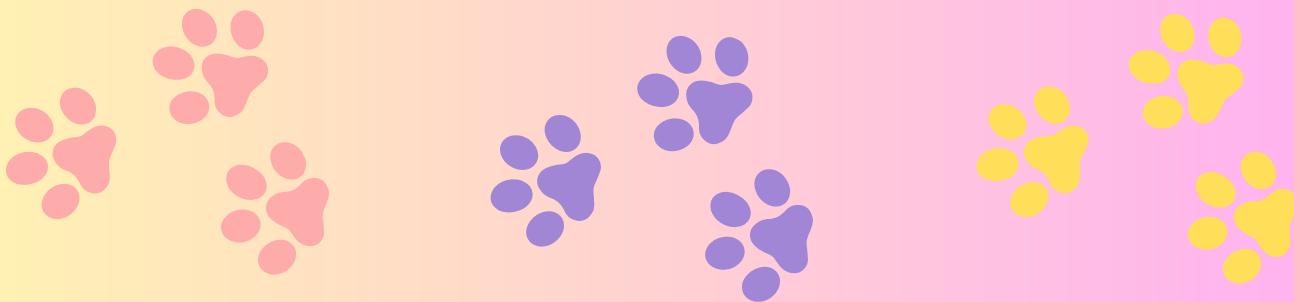
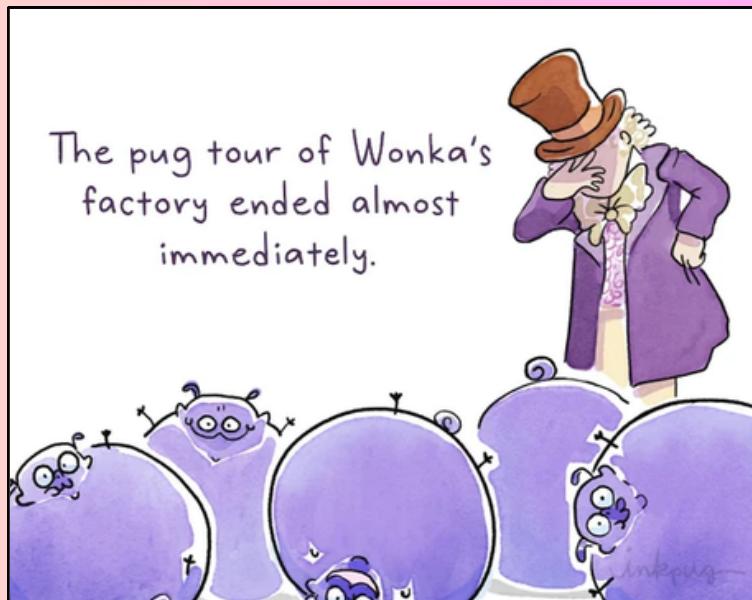
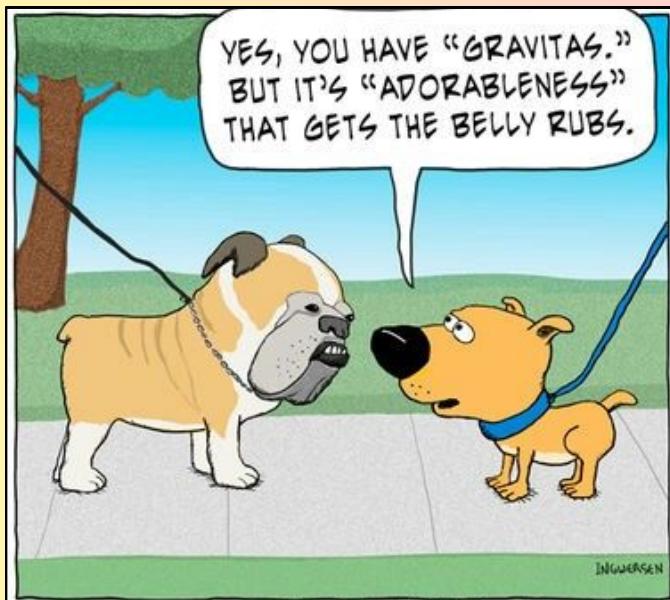
RESULTS

All included dogs were screw-tail brachycephalic breeds with a median age of 6 months. All dogs had ambulatory paraparesis and ataxia, and in 1 dog, signs of spinal hyperesthesia could be elicited. Nonsurgical treatments consisted of restricted exercise without ($n = 5$) or with (3) physiotherapy, physiotherapy without restricted exercise (3), and no exercise modification (2). Seven dogs received additional nonsurgical treatment consisting of prednisolone ($n = 5$) or gabapentin (2). Four dogs were eventually euthanized because of progressive neurologic deterioration, 2 underwent surgery for the same reason, and the remaining 7 dogs survived for ≥ 170 days after diagnosis, despite progressive neurologic deterioration.

CONCLUSIONS AND CLINICAL RELEVANCE

Nonsurgical treatment of congenital thoracic vertebral body malformations was associated with an unfavorable outcome in this group of dogs. Despite this treatment, all dogs had progression of clinical signs. (*J Am Vet Med Assoc* 2018;253:768–773)

Well, we won't learn anything about conservative management for thoracic vertebral malformations from this paper!!! Insufficient numbers. No description or standardization of 'physiotherapy'. Hydrotherapy and physiotherapy were classified as the same thing. All in all, I would say that this paper doesn't tell us anything about whether conservative management works and/or what might or might not work in regard to non-surgical care.



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