Clinical approach to the adult Doberman Pinschers with cervical spondylomyelopathy ('wobbler syndrome')

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SUMMARY

Disk-associated cervical spondylomyelopathy (DA-CSM) is the most prevalent and most typical form of cervical spondylomyelopathy (CSM) in dogs. It is typically seen in the middle-aged Doberman Pinscher. In DA-CSM, caudal cervical spinal cord compression is caused by protrusion of one or more intervertebral disks, sometimes seen in combination with ligamentum flavum hypertrophy and vertebral abnormalities. Several factors have been suggested, but the underlying cause of this multifactorial syndrome is currently unknown. Although clinical signs can vary from cervical hyperaesthesia to non-ambulatory tetraparesis, the most common clinical presentation is a rather characteristic gait disturbance. The diagnosis can be made using myelography, postmyelographic computed tomography (CT-m) or magnetic resonance imaging (MRI). Due to its safe and non-invasive nature, MRI is becoming the imaging modality of choice to diagnose DA-CSM in dogs. There is a lot of controversy concerning the treatment of this disease. Many surgical procedures have been developed, but the ideal surgical technique does not yet exist. All reported surgical techniques carry the risk of significant morbidity. The most common complication after surgery is recurrence of clinical signs after initial recovery. Prognosis for medical management is guarded. Insufficient data is available on objective prognostic indicators that could assist in selecting the appropriate treatment modality for DA-CSM in dogs.

INTRODUCTION

Cervical spondylomyelopathy (CSM) or 'Wobbler syndrome' refers to a collection of disorders in which abnormalities of the cervical vertebrae and surrounding soft tissues (i.e. intervertebral discs and ligaments) result in progressive spinal cord compression. This incompletely understood, multifactorial syndrome typically affects large and giant breed dogs [1-3]. The Doberman pinscher and Great Dane are overrepresented in several studies [4-7]. This syndrome results in clinical signs of ataxia, varying degrees of paresis and cervical hyperaesthesia [1-2]. A large variety of lesions have been attributed to cervical spondylomyelopathy and more than 10 synonyms are found in the literature [8-15]. Over years more separate syndromes have been recognised, based on the characteristic clinical presentation and imaging findings of dogs with CSM [16,17]. More specifically, it has recently been suggested that Doberman Pinschers and Great Danes with CSM could suffer from separate disease entities [3].

The most typical and predominant form of CSM is disk-associated cervical spondylomyelopathy (DA-CSM), also commonly referred to as disk-associated wobbler syndrome [16]. This specific type of CSM is typically seen in middle-aged large breed dogs, in particular the adult Dobermann Pinscher. In DA-CSM, cervical spinal cord compression typically results from the protrusion of the intervertebral disk between the sixth and seventh cervical vertebrae (C6-C7) and/or between the fifth and sixth cervical vertebrae (C5-C6). This disk-associated spinal cord compression is sometimes seen in combination with generally mild vertebral malformations and dorsal compression resulting from hypertrophy of the ligamentum flavum [1]. It is not uncommon for dogs with DA-CSM to present with more than one site of caudal cervical spinal cord compression [6,7,17]. Although several factors have been proposed, the underlying cause of DA-CSM remains currently unknown [2,3]. This manuscript deals primarily with DA-CSM and will outline the clinical presentation, diagnosis and treatment options for this specific type of CSM.

CLINICAL PRESENTATION

Animals affected with DA-CSM are usually older than four years of age. Several large breed dogs can be affected, but the Doberman pinscher is overrepresented [4-6]. Clinical signs can vary from cervical hyperaesthesia to non-ambulatory tetraparesis. The most common presentation is a gait disturbance. The pelvic limbs are usually more severely affected than the thoracic limbs. A wide based ataxia with paresis of the pelvic limbs is usually noted [18]. Thoracic limb involvement with a short stilted gait can occur. Affected dogs often show a characteristic 'disconnected' or 'two-engine' gait, with the thoracic and pelvic limbs advancing at different rates. Although a gradual onset is noticed most commonly, clinical signs can also occur or exacerbate more acutely [1,18]. The severity of clinical signs does not always correlate with

the degree of spinal cord compression and outcome after treatment [19]. In dogs with apparently normal thoracic limbs, it is sometimes difficult to distinguish DA-CSM from a thoracolumbar lesion [17]. Cervical hyperaesthesia may be seen but is usually not overtly present: affected dogs typically resist dorsal extension of the neck [20].

DIAGNOSIS

Survey radiography

Survey radiographs may be indicative for the presence of DA-CSM, but they are not conclusive [18]. General anesthesia is usually necessary to obtain correct positioning of the dog. In dogs with DA-CSM, changes can be seen in the vertebral body, the vertebral canal and the intervertebral disk space (Figure 1). The altered shape of the vertebral body can range from varying degrees of loss of the ventrocranial border, to a triangularly shaped vertebral body. The vertebral body can be tilted with its craniodorsal tip into the vertebral canal. Spondylosis deformans may be seen ventral to the intervertebral disk space, with associated changes in the opacity of the vertebral body. Narrowing or collapse of the intervertebral disk space is frequently seen in dogs with DA-CSM [1,21,22]. The vertebral canal may be stenotic and funnel shaped [21-23]. Changes on survey radiographs do not always correlate with evidence of spinal cord compression on more advanced imaging techniques [14,21,24].

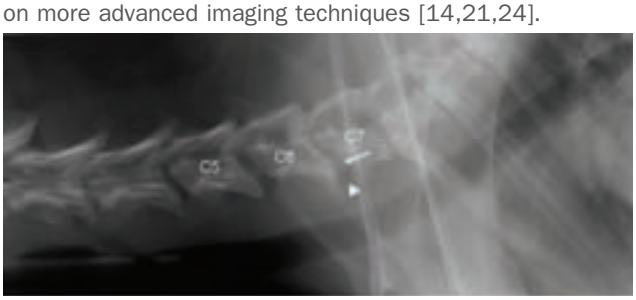

Figure 1. Survey radiograph of a four-year-old Dobermann. Severe narrowing of the intervertebral disk space between C6-C7 (arrow). New bone formation is visible on the ventral aspect of C6-C7 (arrowhead). Malformation of the ventrocranial border of C7.

Some dogs with severe radiographic abnormalities will demonstrate no spinal cord compression or clinical signs [21]. Conversely, survey radiographs may be unremarkable in some dogs with obvious clinical signs of DA-CSM [22].

MYELOGRAPHY

In myelography or contrast radiography, the spinal cord is outlined by a contrast medium injected into the subarachnoid space. For many years, myelography has been the method of choice for diagnosing DA-CSM, but has now largely been replaced by magnetic resonance imaging (MRI) [2,3]. In lateral views, abnormalities are seen in both the ventral and dorsal aspects of the vertebral canal (Figure 2A). Ventral extradural compression related to the intervertebral disk is the most common finding. Multiple sites of compression are common. Dorsal compression caused by hypertrophy of the ligamentum flavum can also be seen in some dogs [22].

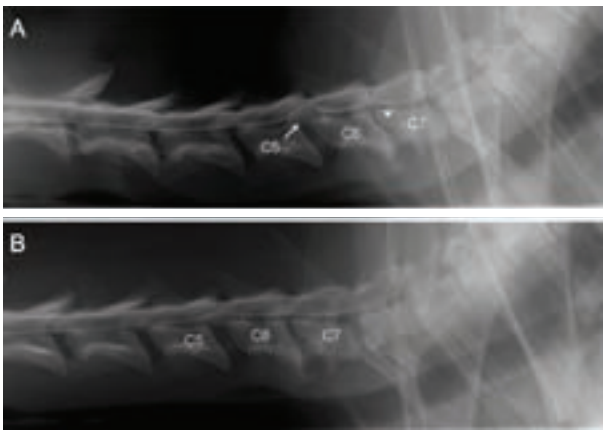

Figure 2. Lateral neutral (A) and traction (B) myelogram of the same dog as in Fig 1. (A) Although the survey radiographs do suggest a compressive lesion between C6 and C7, severe extradural spinal cord compression is noted between C5 and C6 (arrow). A smaller compressive lesion is noted between C6 and C7 (arrowhead). (B) The severity of the compressive lesion reduces remarkably in size after linear traction. This is an example of a traction-responsive lesion.

The merit of applying traction during myelography has been discussed and questioned extensively [3,25]. Lesions may be categorised based on whether or not compression changes in the 'traction' position. Traction views are performed by applying tension to the head in a forward direction and to the forelimbs in a caudal direction. Lesions are termed 'static' when the degree of compression remains the same, whereas 'traction-responsive' lesions improve after performing linear traction [22] (Figure 2B). Traction usually decreases spinal cord compression caused by protruding annulus fibrosus and other ligamentous structures [4]. Therefore most dogs with DA-CSM will show traction-responsive lesions. This subdivision of lesion types can guide the surgeon to decide on the best surgical procedure to perform [18]. Traction-responsive lesions have been suggested to benefit from distraction-stabilisation surgery, while static lesions would benefit from direct decompressive surgery [17]. The degree of compression can also change as the neck is moved between flexed, neutral and gently extended positions. Extension usually exacerbates and flexion usually relieves spinal cord compression in dogs with DA-CSM [17,18]. Performing flexion or extension is not without risk and should be done either with extreme care or not at all [21]. A distinct disadvantage of myelography is its invasive nature [18,22]. Seizures and transient neurological deterioration are the most important complications following myelography [22]. Postmyelographic seizures have been reported in 27% [25,26], while transient neurological deterioration was reported in 14% of dogs with DA-CSM [26].

Computed Tomography and Computed Tomography myelography

Computed tomography (CT) generates successive cross-sectional images with excellent detail, particularly of the bony structures, which can be reconstructed in different planes (Figure 3A). Because of its inability to delineate the spinal cord, conventional CT does not provide as

much information as conventional myelography [27]. When CT is used in combination with a subarachnoidal injection of contrast medium (postmyelographic computed tomography), a good delineation of the spinal cord can also be obtained. An optimal postmyelographic computed tomography (CT-m) image is obtained when a lower dose of contrast medium is used than in a conventional myelographic study [28]. The normal canine cervical spinal cord has a somewhat round appearance and is surrounded by a subarachnoid space of relatively even diameter (Figure 3B). In dogs with DA-CSM the ventral subarachnoid space is attenuated and the spinal cord appears to be displaced from the floor of the vertebral canal (Figure 3C). These abnormalities are caused by the protruding intervertebral disk [27]. Traction studies can also be performed by CT myelography [29]. Although an optimal CT myelography study is performed with a lower dose of contrast medium than a conventional myelographic study [28], this technique still carries the risk of complications such as postmyelographic seizures [7].

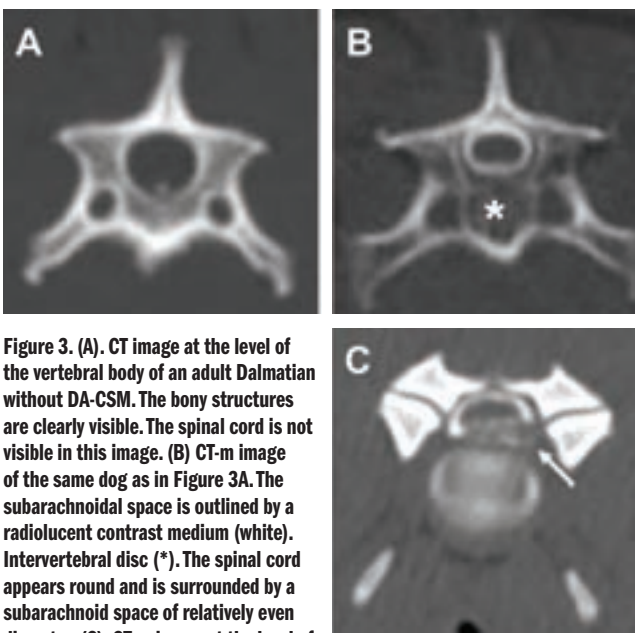

Figure 3. (A). CT image at the level of the vertebral body of an adult Dalmatian without DA-CSM. The bony structures are clearly visible. The spinal cord is not visible in this image. (B) CT-m image of the same dog as in Figure 3A. The subarachnoidal space is outlined by a radiolucent contrast medium (white). Intervertebral disc (*). The spinal cord appears round and is surrounded by a subarachnoid space of relatively even diameter. (C). CT-m image at the level of C5-C6 of the same dog as in Figures 1 and 2. A right-sided intervertebral disk protrusion with spinal cord compression (arrow). The spinal cord has a flattened appearance and the ventral subarachnoid space is attenuated.

Magnetic Resonance Imaging

Magnetic resonance imaging is becoming more and more the imaging modality of choice for the diagnosis of DA-CSM in dogs [2]. MRI allows direct, non-invasive, multiplanar imaging without loss of detail and an excellent soft tissue characterisation with an absence of ionizing radiation [30,31]. A distinct advantage is the ability to directly assess the spinal cord parenchyma without the need for subarachnoidal contrast injections [31]. Therefore, the complications occasionally seen with myelography or CT-m are not seen after MR imaging [18]. Spinal cord compression, intervertebral disk degeneration, intervertebral disk protrusion and spinal cord signal

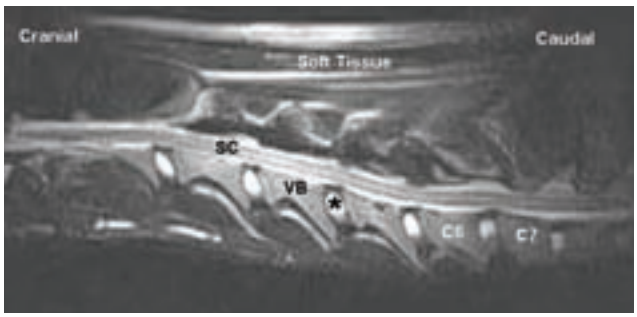

Figure 4. T2-weighted sagittal MR image of an adult Dobermann Pinscher without DA-CSM. The spinal cord (SP) is surrounded by the hyperintense subarachnoidal space (white). The subarachnoidal space is visible at each point and never interrupted. VB = Vertebral body. * = a normally hydrated intervertebral disk. Between C6 and C7: partial intervertebral disk degeneration, characterised as a partial loss of hyperintensity of the disk.

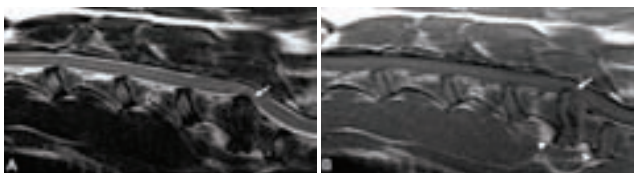

Figure 5. Sagittal T2-weighted (A) and T1-weighted (B) MR images in a seven-year-old Doberman pinscher with ambulatory paraparesis. Left is cranial. Complete degeneration and protrusion intervertebral disk C6-C7. Ventral extradural spinal cord compression (arrow). Abnormal, almost triangular, shaped vertebral body C7. Spondylosis deformans ventral to intervertebral disk space C6-C7 (arrowheads).

changes are abnormalities that can be revealed in dogs with DA-CSM using MRI imaging [32] (Figures 4 and 5). Spinal cord compression can be evaluated on sagittal and transverse T2-weighted images as a loss of hyperintense cerebrospinal fluid (CSF) signal around the spinal cord or as a change in shape of the spinal cord from round to oval on the transverse image [31]. Intervertebral disk degeneration is characterised by a loss of hyperintensity of the disk on T2-weighted images. Abnormal spinal cord signal changes are classified either as hyperintense or as hypointense when they are compared to the normal spinal cord signal intensity adjacent to the abnormal area [32]. Hyperintense T2-weighted signal changes, within the spinal cord, are believed to reflect a broad spectrum of spinal cord abnormalities such as edema, inflammation, ischemia, gliosis and myelomalacia. The exact clinical and prognostic role of spinal cord signal changes is not yet known [33]. Several authors have demonstrated the possibility of performing traction studies during an MRI study [32,34]. A possible disadvantage of MRI in the evaluation of the spine is its high sensitivity. It is not uncommon to identify MRI abnormalities, such as intervertebral disk degeneration and spinal cord compression, in the caudal cervical region of clinically normal, older large-breed dogs [32,35]. Based on MR images alone, it is not always possible to differentiate clinically normal dogs from dogs with DA-CSM [36]. This highlights the importance of evaluating imaging studies in the light of thorough neurologic examination findings. Other disadvantages are the limited availability, and the high costs associated with MRI [18].

TREATMENT

A lot of controversy and discussion exists concerning the treatment of DA-CSM, the role of medical management and the type of surgery that is most likely to give the best results in each individual case [16].

Medical treatment

Disk-associated cervical spondylomyelopathy is often considered a progressive disease in which surgery is necessary to halt progression of clinical signs [17,18]. Only limited information is available about the medical management of this disorder. Recent studies have evaluated the outcome after restricted exercise for several weeks in combination with a tapering course of anti-inflammatory doses of oral corticosteroids [5,6,19]. Unfortunately, medical treatment for DA-CSM was associated with a guarded prognosis. Reported success rates varied between 38% and 45% [6,19] and most dogs suffered from obvious corticosteroid related side effects [19]. If medical treatment of DA-CSM fails, progression of clinical signs is generally rapid, with most dogs being euthanised within the first six months after diagnosis [6,19]. A study, focusing on dogs available for at least six months after a diagnosis of CSM was made, found that 54% of included dogs improved and 26% stayed the same after medical treatment of CSM [5]. Several studies have aimed to identify prognostic indicators for dogs treated medically for DA-CSM [5,6,19]. A higher degree of spinal cord compression on MRI and a more severe degree of neurological deficits one month after diagnosis have been associated with a poor outcome after medical treatment for DA-CSM [19].

Surgical treatment

More than 20 surgical procedures have been described to treat DA-CSM. Although many authors claim good success rates, the large number of reported techniques reflects the difficulty of treating DA-CSM [37-50]. All surgical procedures for the treatment of DA-CSM have a potential for morbidity and postoperative complications [1,18]. There are two basic types of surgery: ventral decompression and vertebral distraction-stabilisation procedures [18]. Suggested factors governing the choice of surgical procedure are the appearance of the spinal cord on imaging, in particular the traction studies, the number of sites of spinal cord compression, the degree of vertebral malformation and the presence of nerve root compression (thoracic limb lameness) [17]. Recently, the clinical use of intervertebral disk arthroplasty has been described and studies are underway to evaluate the efficacy of this new surgical approach in a larger number of patients [50]. Dorsal decompressive surgery by a continuous cervical dorsal laminectomy has been suggested in dogs with DA-CSM that have three or more sites of ventral spinal cord compression [51,52].

Ventral Decompression

Ventral decompression by a standard ventral slot technique is appropriate for single, static lesions [17,18,20]. Ventral decompressive surgery can be challenging for dogs with DA-CSM because of the possibility of vertebral malformations, limited access to the caudal cervical intervertebral disk spaces and intraoperative bleeding due to possible adhesions between the hypertrophied annulus and venous plexus [17,18]. The two main disadvantages of this surgical technique are the inability to perform surgery on two adjacent disk spaces and the inability to treat dorsal compressions due to ligamentum flavum hypertrophy [17.18]. Short-term deterioration is common, even among dogs that have good long-term results [4]. Reported success rates vary from 66% to 100% [4,5,14,37,53,54]. The most common and important complication after ventral decompressive surgery in dogs with DA-CSM is recurrence of clinical signs after initial recovery [18]. It is commonly believed that about 20% to 30% of the dogs undergoing single level decompression suffer a second episode of neurological signs within 2-3 years [4,38]. Most often, the reason for this neurological deterioration is the development of a compressive lesion at an adjacent disk space, which is called ‘adjacent segment disease’ or a ‘domino lesion’ [2,18]. This occurs independently of the surgical technique performed, also after distraction-stabilisation techniques [4,16].

Vertebral distraction-stabilisation

The primary indications for a distraction-stabilisation procedure are the presence of a traction-responsive lesion, dorsal spinal cord compression and the presence of nerve root compression [17,18]. Many different techniques have been developed, including vertebral distraction and stabilisation with vertebral body pins or screws and bone cement [38], a screw and washer [39], a screw and double washer [40], interbody bone cement plug [40], position screw technique [42], spinal locking plates [44,47], spinal locking plates in combination with a cortical ring allograft [45] or an intervertebral cage [49], a distractable intervertebral fusion cage [29]. All of these different surgical techniques are based on the same principle. A ventral slot defect is drilled to a depth of three-quarters of the height of the intervertebral disk space. In this way the dorsal annulus is preserved and the vertebral canal is not entered. Traction is applied to the adjacent vertebrae using vertebral distraction instruments. The two vertebrae are then rigidly stabilised with an orthopedic implant to maintain distraction [18]. Linear traction provides immediate cord decompression by stretching the dorsal annulus, dorsal longitudinal ligament and ligamentum flavum. By stabilising the adjacent vertebral bodies the hypertrophied soft-tissue structures are allowed to atrophy with time [1]. The advantage of not entering the vertebral canal is offset partly by the risk of implant failure or other implant-associated complications such as loosening, migration or breaking of implants and vertebral end-plate fracture due to inadequate contact between the orthopedic implant and the vertebral endplate. Implant failure can be asymptomatic in some patients [18]. As in other surgical

techniques, it seems to be very difficult to perform surgery on more than one intervertebral disk space at the same time. Adjacent segments disease occurs with the same incidence with this technique as with ventral decompressive surgery [4,17,18]. Few surgical techniques combine the concepts of direct decompression with distraction-stabilisation at one or two adjacent disk spaces [46,48].

Cervical disk arthroplasty

As stated above, the most common complication after surgical treatment for DA-CSM is adjacent segment disease. Although the exact pathophysiology of this complication is still unknown, it has been hypothesised that fusion or distraction of an intervertebral disk space will alter the biomechanics of the adjacent spinal segments [55]. These altered biomechanics may cause or contribute to the development of adjacent segment disease [56]. Disk arthroplasty has been evaluated in a canine cadaveric study [57] and more recently on a limited number of dogs with DA-CSM [50]. Cervical disk arthroplasty aims to reestablish the normal intervertebral disk space, while preserving the motion pattern of the affected intervertebral segment [50,57].

PROGNOSIS

Several surgical studies report success rates between 70% and 80% immediately postoperative [38-43, 45]. Conversely, reported long-term mortality rates vary from 19% to 43% [4,40,41]. After both ventral decompressive and vertebral distraction-stabilisation techniques, a second episode of clinical signs is seen in about 20% to 30% of the cases [4,16-18]. Although the underlying pathophysiology of adjacent segment disease is currently unknown, this is possibly related to altered biomechanics after spinal fusion, a natural progression of a multifocal disease or a combination of both [16]. It is difficult to compare the results of different surgical techniques, due to the differences in case selection, in the definition of a successful outcome and in the length and descriptions of follow-up [16]. Some surgical reports deal with 'CSM in general' and do not focus on DA-CSM in particular. Suggested prognostic factors for surgical treatment include, the number of compressive lesions, the severity of neurological deficits, the duration of clinical signs and the degree of spinal cord compression [16-18,20]. In some cases, surgery will only halt the progression of the disease. In these cases, irreversible spinal cord damage has probably occurred. Several authors suggest the use of advanced medical imaging, such as CT-m and MRI to diagnose spinal cord atrophy [7,17,27]. A possible correlation between the diagnosis of suspected spinal cord atrophy and a poor outcome has not yet been investigated in veterinary medicine. Prognosis after medical management for DA-CSM is guarded (see above). Suggested prognostic indicators for medical treatment of DA-CSM include type of clinical signs, the degree of spinal cord compression and clinical evolution one month after diagnosis of DA-CSM [6,19]

CONCLUSION

Disc associated cervical spondylomyelopathy is a relatively common cause of chronic cervical spinal cord compression in large breed dogs. This disease can be very challenging, both for the referring veterinarian and even for the specialist. One of the key problems is the discrimination between this and other types of CSM. The advantages and disadvantages of the different diagnostic procedures should be considered when approaching a dog suspected to have DA-CSM. Although considered a surgical disease, the ideal surgical procedure still does not exist. In addition, there is little data available on prognostic factors for this disease.

References are available at www.veterinaryirelandjournal.com

Reader Questions and Answers

1. DA-CSM IS TYPICALLY ASSOCIATED WITH:
a) Hypertrophy and malformation of the articular process joints
b) Dorsal vertebral arch hypertrophy and malformation
c) Severe vertebral canal stenosis, causing osseous spinal cord compression
d) Rather mild malformations of the ventrocranial border of the vertebral body

2. THE MOST COMMON CLINICAL PRESENTATION OF DOGS WITH DA-CSM IS:
a) Severe cervical hyperaesthesia
b) Ambulatory paresis and ataxia, predominantly affecting the thoracic limbs
c) Ambulatory paresis and ataxia, predominantly affecting the pelvic limbs
d) Non-ambulatory tetraparesis

3. THE MOST COMMON COMPLICATION AFTER MYELOGRAPHY IN DOGS WITH DA-CSM IS:
a) Transient neurological deterioration
b) Postmyelographic seizures
c) Anaphylactic adverse reaction
d) Bradycardia

4. WHICH OF THE FOLLOWING IS AN IMPORTANT ADVANTAGE OF MAGNETIC RESONANCE IMAGING
a) It is widely available and relatively cheap
b) It gives superior resolution of bony structures
c) It is a safe and non-invasive procedure
d) It is reliable to differentiate clinically normal dogs from dogs with DA-CSM (i.e. very good sensitivity and specificity)

5. MEDICAL TREATMENT OF DA-CSM:
a) Is associated with a guarded outcome
b) Is associated with an excellent outcome
c) Gives better results in dogs with a short duration of clinical signs
d) Is not associated with any of the statements above

6. THE MOST IMPORTANT SURGICAL COMPLICATION IN DOGS WITH DA-CSM IS:
a) Intra-operative haemorrhage
b) Iatrogenic spinal cord damage
c) Implant failure
d) Recurrence of clinical signs after initial recovery

ANSWERS: 1. D, 2. C, 3. B, 4. C, 5. A, 6.D.