New considerations about Chiari-like malformation, syringomyelia and their management

Background: Chiari-like malformation (CM) is a developmental condition, characterised by a conformational change and overcrowding of the brain and cranial cervical spinal cord. CM-associated pain (CM-P) and syringomyelia are increasingly being diagnosed, due to the rising popularity of predisposed brachycephalic breeds and the availability of MRI in veterinary practices.

Aim of the article: This article aims to update the veterinary profession on these conditions, and provides a guide to diagnosis and treatment of clinically relevant disease.

Introduction
Chiari-like malformation (CM) is a complex developmental condition of the skull and craniovertebral junctions, and is characterised by a conformational change and overcrowding of the brain and cervical spinal cord, particularly at the craniospinal junction. Obstruction to cerebrospinal fluid (CSF) channels can result in pain and a tendency for fluid cavitation of the spinal cord, called syringomyelia (SM) (Figs 1 to 3). The fluid within the cavities (singular syrinx, plural syringes) is similar to CSF. These fluid pockets can expand and can cause irreversible damage to the spinal cord, resulting in clinical signs of pain and neurological deficits.

The rising popularity of predisposed brachycephalic toy breed dogs and availability of MRI has seen CM-associated pain (CM-P) and SM becoming a more common diagnosis in veterinary medicine. CM-P can be a challenging diagnosis because the signs of pain are non-specific and, unlike SM, MRI diagnosis of CM-P is poorly defined or ambiguous. A degree of CM is ubiquitous in predisposed breeds and CM-P can be late onset, meaning it is challenging to distinguish clinically affected dogs using MRI (Figs 1, 2).

Nomenclature
The eponymous term Chiari malformation refers to the first detailed description of the analogous human condition by Hans Chiari in 1891 and classically describes a cerebellar herniation through the foramen magnum. Alternative names for the canine condition include caudal occipital malformation syndrome (COMS) and occipital hypoplasia. None of these names are ideal because the malformation is more complex than a cerebellar herniation, simple occipital bone abnormality or small volume caudal fossa.

An alternative term, brachycephalic obstructive cerebrospinal fluid channel syndrome (BOCCS), was proposed to reflect the connection to brachycephaly and obstruction of CSF pathways. However, as CM was the terminology decided at a round table discussion (Capello and Rusbridge 2007), this is the most commonly accepted moniker.

The nomenclature of SM has morphed over the years since the first description in the early 19th century. Authoritative sources use SM rather than historical terms syringohydromyelia, hydrosyringomyelia or hydromyelia. This is because the anatomical distinction between these terms is theoretical rather than a reality (Rusbridge and Flint 2014). It is conventional in veterinary medicine to refer to a central syrinx, less than 2 mm in transverse diameter, as a central canal dilation (Fig 3), even though the ependymal lining of the central canal is disrupted with only minor dilation.
Companion animals

(Radojcic and others 2007). Non-inflammatory spinal cord oedema, as distinct from cavities containing free fluid, is referred to as presyrinx (presyringomyelia). Presyrinx most commonly affects the dorsal and ventral columns of the spinal cord and may eventually progress to SM (Fig 3). The oedema can reverse if the cause can be addressed. CNS inflammatory diseases can also cause spinal cord oedema and are alternative differentials for spinal cord oedema (Fig 4).

**Signalment and prevalence**

Brachycephaly with airobrhythmy (i.e., retroflexion of the facial skeleton on the cranial base) is the major risk factor for CM-P and SM. Any animal with skull shortening including the muzzle with, in dogs, concurrent whole body miniaturisation could be predisposed. The condition is most common in Cavalier King Charles spaniels (CKCS) and their crosses (e.g., cavapoos), especially if the cross is with a smaller dog than a CKCS. Other breeds with high

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**Fig 1:** Spectrum of Chiari-like malformation (CM) and syringomyelia (SM) in Cavalier King Charles spaniels (CKCS), ranging from clinically unaffected to affected.

(a) Normal CKCS (CM-N). Three-year-old male, neutered, 17 kg dog that presented with lumbar pain due to L3/L4 intervertebral disc (IVD) extrusion. (b) Normal CKCS (CM-N). Seven-year-old male, entire, 16 kg dog illustrates the grey area between normal and abnormal. This dog presented with acute-onset cervical pain related to a C3/C4 IVD extrusion. There was no previous history consistent with CM-associated pain (CM-P) and the cervical pain resolved after surgical decompression of the IVD extrusion. The appearance of the skull, craniovertebral junction and brain is only subtly different from the clinically affected dogs and there is a cerebellar herniation. (c) CKCS with CM-P. Seven-year-old female, neutered, 11 kg dog was presented because of repeated vocalisation as if it was in pain when falling asleep, together with unwillingness to rise, an aversion to its ears being touched, walking slowly upstairs, and yelping when being picked up from under the sternum. (d) CKCS with SM-Specific signs (SM-S). Seven-year-old female, neutered, 7.3 kg dog that presented with a scratching action towards the shoulder area without making skin contact and when walking on a lead, mostly with the right pelvic limb but sometimes with the left. Red line – outline of the muzzle, stop and ‘forehead’. The angle of stop and the size of frontal sinus progressively reduces with clinical affectedness. Yellow line – progressive rostral flattening of the forebrain with clinical affectedness. Green shading – progressive reduction and ventral displacement of the olfactory bulbs with clinical affectedness. Purple shading – progressive shortening and reduction of the basilarium, especially the presphenoid bone with clinical affectedness. Turquoise arrows – in CM-P and SM-S the forebrain is displaced caudally. The space for the hindbrain is compromised rostrally by the forebrain and caudally by the small caudal skull. The cerebellum loses its rounded shape and is pushed out of the foramen magnum. Blue line – with increasing clinical affectedness, the supraoccipital bone becomes flatter and shorter and the opisthion (dorsal foramen magnum) becomes rostral with respect to the occipital crest. Orange star – progressive reduction in bony tissue of the occipital crest as the dog becomes more clinically affected; however, sexual dimorphism may also influence this

**Fig 2:** Spectrum of changes of Chiari-like malformation (CM) and syringomyelia (SM), ranging from clinically unaffected to affected Cavalier King Charles spaniels (CKCS). T2W MRI of the midsagittal head and cranial cervical region.

(a) Normal CKCS (CM-N). (b) Normal CKCS (CM-N), illustrating the grey area between normal and abnormal. (c) CKCS with CM-associated pain (CM-P). (d) CKCS with SM-Specific signs (SM-S). The signalment and descriptions of dogs are the same as for Fig 1. Pink shading – from (a) to (d), the rough outline of the forebrain changes from a rugby to a football shape because of increasing brachycephaly. White line – progressive ventral rotation of the axis of the brain. Yellow star – SM. Turquoise line – change in the conformation of the craniospinal junction because of craniovertebral junction (occiput, atlas, and axis) malformation. The atlas is closer to the skull with cervical flexure and acute angulation of the odontoid peg, resulting in kinking or elevation of the neuroparenchyma.
Companion animals

prevalence include King Charles spaniels, griffon bruxellois, affenpinschers, Chihuahuas, Yorkshire terriers, maltese, Pomeranians and brachycephalic cats, especially Persians. Occasionally affected dog breeds include French bulldogs, Boston terriers, pugs, havanese, miniature dachshunds, miniature/toy poodles, bichon frisé, and miniature pinschers.

Surprisingly, many toy breeds with extreme facial foreshortening such as the Pekinese, Japanese chin and shi tzu, are not as predisposed, perhaps reflecting a different skull shape, brain size and genetic heritage.

When MRI is performed in predisposed breeds, CM is commonly reported, and SM may be an incidental finding. Care must be taken to not overdiagnose, and MRI results should be related to historical and clinical findings. Cerda-Gonzalez and others (2009) found that 92 per cent of CKCS had at least one cranioceval morphological abnormality detected on MRI, and studies looking at groups of 16 or more clinically unaffected CKCS found a high incidence of SM, ranging from 26.5 per cent (Cerda-Gonzalez and others 2009) to 65.4 per cent (Rusbridge and others 2007). These figures increased to 42 per cent and 74.5 per cent, respectively, when dogs with clinical signs were added to the population.

Dogs may be presented with the disease at any age, although many dogs (approximately 45 per cent) will develop first signs of the disease within the first year of life, and approximately 40 per cent of cases have first signs between one and four years of age. As many as 15 per cent develop signs as mature dogs (aged between six and eight years of age) (Plessas and others 2012, Thofner and others 2015).

Pathophysiology

Chiari-like malformation

CM is a developmental malformation characterised by neuparenchymal disproportion – the ‘box’ (skull and cranial cervical vertebrae) is too ‘short’ for the contents (brain and cranial cervical spinal cord). The key feature of canine CM is premature suture closure (craniosynostosis) and insufficiency of bones forming the skull base and caudal skull (Figs 1, 2). CM is also characterised by a reduction in craniofacial tissue with a loss of the frontal sinus and a more defined ‘stop’ (Knowler and others 2020). The ‘stop’ is the pronounced angle between the nasal/maxilla bones and the frontal bones, which is a defining feature of domesticated mesaticephalic and brachycephalic dogs and by contrast is not present in wolves. The skull

Fig 3: T2W midsagittal cervical MRI (left) and T2W transverse MRI at level of C2/C3 (right), showing the spectrum of changes from normal to syringomyelia (SM). (a) Normal MRI appearance in a dog. (b) Central canal dilatation (blue arrow). (c) Area of presyrinx (green arrow) in the spinal cord (dorsal) and ventral columns surrounding a small syrinx. (d) The same dog as in (c) but six years later and now with a fully developed SM (red arrows) which is asymmetrical and expanding the spinal cord

Fig 4: MRI images from a three-year-old male, neutered Cavalier King Charles spaniel (CKCS) presenting with cervical pain and reluctance to walk. (a) T2W midsagittal MRI of neuroaxis from hindbrain to T8. (b) Transverse T2W MRI at level of C2. (c) Parasagittal T2W MRI at the level of the mesial temporal lobe. (d) Transverse T2 MRI at the level of the mesial temporal lobe and thalamus. (e) Transverse fluid-attenuated inversion recovery (FLAIR) MRI at the level of the mesial temporal lobe and thalamus. (f) Transverse fluid-attenuated inversion recovery (FLAIR) MRI with paramagnetic contrast at level of C2. (g) Midsagittal T1W MRI with paramagnetic contrast of neuroaxis between hindbrain and C3. Hyperintensity on T2W images suggests spinal cord oedema (turquoise arrow), but unlike the case in Fig 3 the oedema is within in the grey matter rather than the dorsal columns. The white matter of the dorsal column is hypointense on T2W (red star) and enhances with paramagnetic contrast (red arrows). These images support suspicion of multifocal inflammatory or neoplastic disease with hyperintensity in the region of the mesial temporal (green arrows) and periventricular and meningeal tissue – meningoencephalomyelitis is an important differential for spinal cord oedema. CSF analysis revealed a mononuclear (lymphocyte dominant) pleocytosis with a white blood cell count of 1386 cells/ul and a protein concentration of 614.8 mg/dl. PCR for antigen receptor gene rearrangements clonality did not support lymphoma. Testing for common infectious diseases was negative
insufficiency results in rostrotentorial (forebrain) crowding which further reduces the functional caudotentorial space and causes hindbrain herniation. In addition, some predisposed breeds, such as CKCS, have comparatively big brains.

**Chiari malformation-associated pain**

CM-P is defined as the clinical signs of pain relating to CM. Compared to clinically normal dogs with CM (CM-N), dogs with CM-P have more extreme brachycephaly; that is, shorter cranial base, more craniofacial hypoplasia with greater neuroparenchymal disproportion and overcrowding (Knowler and others 2017) (Figs 1, 2).

**Syringomyelia**

There is yet to be an entirely satisfactory explanation of how fluid cavities develop in the spinal cord following CSF pathway obstruction. Whether syrinx fluid is derived from extracellular fluid or CSF is

| Clinical signs                              | Frequency (% affected dogs) | Notes                                                                 |
|---------------------------------------------|-----------------------------|----------------------------------------------------------------------|
| Vocalisation                                | 65                          | Yelping described by the owner as 'out of nowhere', spontaneous or when the dog is moving, while lying resting or asleep, when lifted under the sternum, or on rising |
| Spinal pain                                 | 55                          | Hyperaesthetic to palpation in the cervical, thoracolumbar or caudal lumbar / lumbosacral region |
| Activity change                             | 38                          | Described as exercise intolerant, unwilling to exercise, lethargic or sleeping more |
| Refusal/hesitation/difficulty climbing stairs or jumping | 35                          | Described as refusing/hesitating/difficulty or vocalisation when jumping or climbing stairs |
| Scratching or rubbing of the head or ears   | 28                          | Skin and ear disease should be excluded |
| Aversion to the ears/head/neck being touched or groomed | 25                          | Owner reports that the dog has an aversion to touch of this body part, yet tolerates touch or grooming elsewhere |
| Sleep disruption                             | 22                          | Described as being restless in the night or having disturbed sleep |
| Timid/anxious                               | 14                          | Change in behaviour described by owner |
| Withdrawn                                    | 13                          | Change in demeanour described by owner |
| Forelimb hypermetria                        | 10                          | 'Goose stepping'-type gait characterised by increased proximal joint movement giving a tendency to overshoot. May also have decreased distal joint movement due to increased muscle tone (due to cerebellar dysfunction or a spinocerebellar tract lesion) |
| Aggression                                  | 8                           | Change in behaviour or uncharacteristic belligerence to other dogs/people |
| Abnormal head/neck posture when awake       | 8                           | Head held down or reluctant to move neck |
| Vocalisation when greeting, refusal to rise to greet, or no longer greeting | 5                           | Yelping during greeting or refusal to get up and greet owner |
| Sleeping with an elevated or unusual head posture | 5                           | Brachycephalic obstructive airway syndrome could be an alternate explanation for this sign |
| Aversion to the sternum or flank being touched or groomed | 5                           | Owner reports that the dog has an aversion to touch of this body part, yet tolerates touch or grooming elsewhere |
| Squinting/avoiding light                    | 5                           | Described by the owner as avoiding looking at lights or closing eye in light. Schirmer tear test should be performed to eliminate a diagnosis of keratoconjunctivitis sicca |
| Vocalisation when scratching                | 5                           | Yelping while scratching |
| Licking limb/paw                            | 4                           | Dog licks without evidence of skin or orthopaedic disease and does not respond to trial management for allergic skin disease |
| Pain face                                   | 3                           | Owner describes a grimacing facial expression, suggesting the dog is in pain |
| Aversion to the limb/paw being touched or groomed | 3                           | Owner reports that the dog has an aversion to touch of this body part, yet tolerates touch or grooming elsewhere |
| Vocalisation when defecating                | 2                           | Yelping while defecating |
| Repetitive barking                          | 2                           | Behaviour thought to reflect anxiety; not necessarily due to Chiari-like malformation-associated pain (CM-P) |

Frequency is based on owner-reported and clinical examination findings in CM-P and syringomyelia-affected Cavalier King Charles spaniels (Rusbridge and others 2019)
Companion animals

Table 2: Clinical signs of clinically relevant syringomyelia*

| Clinical signs            | Frequency (% affected with CM-P and SM) | Frequency (% affected with SM-S) | Notes                                                                                                                                 |
|----------------------------|----------------------------------------|----------------------------------|-------------------------------------------------------------------------------------------------------------------------------------|
| Phantom scratching         | 43                                     | 67                               | Rhythmic scratching action towards neck, but not contacting the skin, together with a curvature of the body and neck towards the foot. Induced by light rubbing of the neck or ear region and triggered by excitement, anxiety and exercise. Associated with a large mid-cervical syringe extending to the superficial dorsal horn ipsilateral to the scratching |
| Scoliosis / cervicothoracic torticollis | 18                                     | 27                               | Corkscrew deviation of neck associated with a large mid-cervical syringe extending to the superficial dorsal horn ipsilateral to lateral shoulder deviation and contralateral to the ventral head tilt |
| Weakness                   | 25                                     | 39                               | Thoracic limb and paraspinal muscle weakness associated with a large cervicothoracic syringe                                          |
| Thoracic limb muscle atrophy | 4                                      | 6                                | Thoracic limb muscle atrophy associated with a large C5-T1 spinal segment syringe                                               |
| Postural responses decreased | 10                                     | 15                               | As demonstrated by ‘hopping’, ‘hemi-walking’ and ‘correction of knuckled over paw’ testing. Thoracic limbs are typically more affected than pelvic limbs. Consider other differentials if the dog presents with severe paraparesis |

CM-P: Chiari-like malformation-associated pain; SM-S: Syringomyelia-specific signs
Frequency is based on owner-reported and clinical examination findings in CM-P and SM-affected Cavalier King Charles spaniels (Rusbridge and others 2019)
* That is, SM-S (maximum transverse width equal to or greater than 4 mm)

also controversial. The most accepted theory of the pathogenesis of SM is that obstruction to CSF flow in the subarachnoid space results in a mismatch in timing between the spinal arterial pulse peak pressure and CSF pulse peak pressure. Earlier arrival of peak CSF pressure encourages flow of CSF into the perivascular space. The perivascular space changes in size during the cardiac cycle and is widest when spinal arterial pressure is low. If at that time peak CSF pressure is high, then the perivascular space could act as a ‘leaky’ one-way valve. From the perivascular space, fluid flows into the central canal ultimately resulting in a syrinx (Stoodley 2014).

SM can occur due to any obstruction to CSF pathways and has been reported in a variety of disorders, including acquired cerebellar herniation secondary to intracranial masses, spinal arachnoid diverticulum, spinal cord tethering and inflammatory conditions, such as feline infectious peritonitis. However, in canine medicine by far the most common cause is CM. The pathogenesis of SM associated with CM is predisposed by two morphological phenotypes: more extreme brachycephaly, as for CM-P, and cranio-cervical junction deformation, including changes in angulation of the dens and increased proximity of the atlas to the skull and loss of the cisterna magna (Figs 1, 2). Loss of the cisterna magna or other alterations in the CSF volume will affect the compliance of the CNS. This may be influenced by poor venous drainage, intracranial hypertension, and conformational features of the spinal canal.

Clinical signs

Chiari-like malformation

CM-P is thought to relate to a failure to equilibrate intracranial pressure due to obstruction of CSF pathways. Intracranial pressure is affected by the systolic pulse, venous drainage, the balance between CSF production and absorption and microgravitational effects (eg, when being lifted rapidly or when the head position moves rapidly).

In people, pain in CM is exacerbated by the Valsalva manoeuvre, a brief increase in intrathoracic pressure, for example when coughing or with abdominal straining. Signs of canine CM-P (Table 1) include:

- Vocalisation (described as without obvious trigger, when shifting position when recumbent and when being lifted under the sternum to a height);
- Spinal pain;
- Head and ear rubbing or scratching;
- Aversion to touch;
- Refusal or difficulty jumping or climbing stairs;
- Exercise intolerance/reduced activity;
- Sleep disruption;
- Behavioural change described as becoming more anxious, timid, aggressive or withdrawn.

Syringomyelia

SM-specific signs (SM-S) are associated with large syringes (in CKCS a maximum transverse width of equal to or greater than 4 mm) and include phantom scratching, scoliosis and sensory and motor signs. The neurolocalisation is consistent with the syrinx location (Table 2).

The signs of CM-P and SM (except phantom scratching) are non-specific. Therefore, other possible explanations should be eliminated. Neurological deficits associated with SM will have the same neurolocalisation as the syringe; however, gait disturbances and paresis may be surprisingly mild even with wide SM involving the entire cervical to lumbar spinal cord. Typically, the thoracic limbs are weaker than the pelvic limbs, reflecting the central spinal cord damage. Other differentials
## Table 3: Common differential diagnosis for CM-P and SM and unrelated comorbidities common in CKCS

| Condition | Clinical signs | Notes |
|-----------|----------------|-------|
| **Other neurological signs** | | |
| Idiopathic epilepsy | Seizures, normal interictal neurological examination | No link between CM/SM and epilepsy has been proved. Considered separate diseases for management purposes |
| Paroxysmal dyskinesia (epileptoid cramping syndrome/Spike's disease) | Episodes of abnormal movement that are self-limiting with long periods of normality between episodes. Abnormal movements may include tremor, alternate limb lifting/flexion and weaving head movements with variable gastrointestinal signs. Animal remains conscious and able to respond to owner (eg, may try to walk to owner) | Possible connection to gastrointestinal dysfunction and gluten sensitivity. Episodes may reduce with a hypoallergenic or gluten free diet |
| Fly catching disorder | Behaviour where the dog acts as if watching/catching a fly (although ignores actual flies). Some may also behave as if their ears or feet are irritated and some can also tail chase. May be more likely with certain light intensities and emotional states | Possible connection to gastrointestinal dysfunction. Episodes may reduce with a hypoallergenic or low protein diet. Neurochemical imbalance has been suggested as some dogs respond to anti-epilepsy drugs or selective serotonin re-uptake inhibitors |
| Myoclonus | Brief jerking of the head, often with buckling of a thoracic limb when the dog is standing or sitting | Older CKCS (over five years old). Initially relatively benign but can be progressive over years and the myoclonic jerks can cause the dog to fall or stumble |
| Episodic falling (paroxysmal exercise-induced dyskinesia) | Falling episodes induced by physical activity, stress and excitement, and manifest with hypertonicity of the limbs resulting in inability to move or even complete collapse. In contrast to epileptic seizures, consciousness is unaffected | Movement disorder associated with BCAN (brevican) mutation. Typically manifests between the age of four months and four years. A genetic test is available |
| Idiopathic facial paresis | Inability to close the eye or move the lips, ears or other facial muscles. Facial sensation (trigeminal nerve) is normal | May be bilateral (not necessarily simultaneously) in CKCS, or associated with vestibular signs |
| **Other causes of head tilt** | | |
| Idiopathic vestibular disease | Acute onset vestibular signs (head tilt, nystagmus, positional strabismus, asymmetrical ataxia) with no proprioceptive deficits or weakness | In CKCS idiopathic vestibular disease has two variations: one in geriatric dogs and one in middle-aged dogs often in conjunction with idiopathic facial paresis |
| Rostral cerebellar artery infarction | Acute-onset paradoxical vestibular signs with other signs suggesting a cerebellar location (menace response deficit, intention tremor, decerebellate posture) | Often improves with supportive care |
| **Other causes of spinal pain or myelopathy (proprioceptive deficits and paresis)** | | |
| Atlantoaxial subluxation | Myelopathy localising to the cranial cervical spinal cord (tetraparesis, increased muscle tone, proprioceptive deficits and possible respiratory compromise) | May be seen in association with CM-P and SM, especially in miniature breeds such as Chihuahuas and Yorkshire terriers |
| IVDD – cervical | Neck pain and variable limb postural deficits. May be associated with neck muscle myoclonus (C3 nerve root compression/irritation) or lameness (C5-C8 nerve root compression/irritation) | In contrast to CM/SM, IVDD typically occurs in older dogs (two years of age or older) and spinal pain is typically acute onset and focal |
| IVDD – thoracolumbar | Acute-onset focal pain in T11-L4 region with variable myelopathy (paraparesis and proprioceptive deficits) | |
| IVDD – lumbosacral | Refusal/hesitation/difficulty in jumping or climbing the stairs. Focal pain in lumbosacral region. Unilateral or bilateral pelvic limb lameness. Hyperaesthesia in the distribution of the L7 nerve root | |
| Meningoencephalomyelitis of unknown origin | Spinal pain with variable neurological signs relating to immune-mediated inflammation of the brain or spinal cord | Inflammatory disease will also result in a high intrathecal signal on T2 weighted images (see Fig 4) |
| Degenerative myelopathy associated with gain of function SOD-1 mutation | Pelvic limb proprioceptive deficits and paraparesis (T3-L3 localisation), progressing over months to paraplegia with faecal and urinary incontinence, and over years to tetraplegia. Non-painful | Even with wide SM, significant pelvic limb weakness and proprioceptive deficits is unusual and other differentials should be considered. A genetic test for the SOD-1 mutation is available |

CKCS: Cavalier King Charles spaniel, CM-P: Chiari-like malformation-associated pain, IVDD: Intervertebral disc disease, SM: Syringomyelia, SOD1: Superoxide dismutase 1
Table 3 continued: Common differential diagnosis for CM-P and SM and unrelated comorbidities common in CKCS

| Condition | Clinical signs | Notes |
|-----------|----------------|-------|
| Otitis media with effusion (OME; also known as primary secretary otitis media) | Sequel of brachycephalic conformation and poor drainage of the middle ear. May cause hearing loss or ear discomfort OME is a possible differential for ear pain or rubbing (Guerin and others 2015); however, Rusbridge and others (2019) did not find OME presence significantly associated with head/ear rubbing or scratching | |
| Allergic skin disease | Generalised pruritus, especially of the feet and abdomen CM-P and SM are not associated with pruritus of the ventral abdomen, feet or tail head | |
| Periodontal disease | Oral pain with inflammation of the gingiva and deterioration of the bone and soft tissue structures supporting the teeth Higher prevalence in small breeds and significant association with cardiac disease (Pereira dos Santos and others 2019) | |
| Other causes of sleep disruption, exercise intolerance or lethargy | | |
| Brachycephalic obstructive airway disease | Exercise, heat intolerance, sleep disordered breathing or sleep apnoea Important differential for sleep disruption. Consider if the dog is waking up spluttering or coughing | |
| Degenerative mitral valve disease | Left apical systolic murmur, exercise intolerance, coughing, respiratory effort, nocturnal dyspnoea, pulmonary oedema Important differential for exercise intolerance | |
| Chronic pancreatitis | Recurrent anorexia, mild bouts of colitis-like faeces, occasional vomiting, increased borborygmi, mild abdominal pain Differential for signs of lethargy and behavioural changes suggesting pain | |
| Gastro-oesophageal reflux disease | Classic signs like regurgitation not always present/obvious. Discomfort or pain may manifest as repetitive tongue licking or restlessness Differential for behavioural changes suggesting pain | |

CKCS: Cavalier King Charles spaniel; CM-P: Chiari-like malformation associated pain; SM: Syringomyelia

Companion animals

should be considered if there is a non-ambulatory tetraparesis or severe paraparesis (Table 3). Likewise, SM is a spinal cord disease and so would not result in epilepsy, facial nerve paralysis or fly catching disorder. CKCS are predisposed to several neurological conditions and comorbidities (Table 3).

**Diagnosis**

**Chiari-like malformation**

CM-P is a diagnosis of exclusion in a predisposed breed or in a dog presenting with the signs mentioned earlier. MRI remains the only diagnostic test to support suspicion of CM-P. Although the bony changes can be demonstrated by CT, MRI is required to detect any associated SM. When undertaking brain and cervical MRI of a predisposed breed, it is recommended that the dog’s microchip or tattoo number (confirmed by the vet) is included on the DICOM (digital imaging and communications in medicine) images in addition to the Kennel Club registration number, if relevant, even if CM-P or SM is not a differential. This is to permit submission to the BVA/Kennel Club’s Chiari malformation/syringomyelia (CM/SM) scheme should the owner request this following the imaging (BVA 2013). For details of MRI protocols to investigate CM and SM see Rusbridge and others (2018).

MRI changes (Box 1) (Figs 1, 2) can support but not confirm the diagnosis of CM-P. Research techniques such as MRI-based morphological measurements or machine learning have yet to be adapted to the clinic (Spiteri and others 2019).

**Syringomyelia**

MRI is required for diagnosis of SM (Box 2). The finding of SM implies a fluid-filled cavity related to disturbance of CSF flow, spinal cord tethering or intramedullary tumour. The cause of SM should be determined (Rusbridge and others 2018). SM is not an appropriate description for myelomalacia or cystic lesions. SM can be an incidental finding, and when interpreting MRI an assessment should be made as to whether the location and severity of the syrinx would account for the signs. It would be exceptional for SM associated with CM to result in a myelopathy localising to T3-L3 with pelvic limb paresis and proprioceptive deficits and normal thoracic limb function. If this is the neurological localisation then other differentials should be investigated (Table 3).

**Progression and prognosis**

There is paucity of large studies on the progression and long-term outcome of dogs affected by CM-P and SM. Studies in breeding CKCS suggest that the proportion of affected CKCS increase from 25 per cent at one year of age to 60 per cent at three years of age. By five years of age, 70 per cent of the population. At one year to 60 per cent at three years of age. By five years of age, 70 per cent of the population have MRI evidence of SM (Parker and others 2011). Although syrinx width increases over time, the rate of increase is not constant and it is my unproven impression, supported by computer modelling, that for many dogs the syrinx develops relatively rapidly but then remains remarkably unchanged over years having achieved a dynamic equilibrium.
Clinical signs will progress in approximately 75 per cent of dogs and approximately 15 per cent will be euthanased because of CM-P and SM-S. However, despite progressive signs, many dogs with signs of pain and phantom scratching respond to medical management and are considered by their owners to have an acceptable quality of life (Plessas and others 2012). Cervicotorticollis may slowly improve despite persistence of the syrinx. Dogs that are presented with SM-S before three years of age seem to have a poorer prognosis and are more likely to develop severe weakness which is more difficult to treat.

Medical management
Medical management of SM is based mostly on anecdotal reports and typically relies on adjuvant analgesics and other unlicensed medication (Fig 5) (Table 4). There have been three short-term (14 to 25 day) clinical trials assessing carprofen, gabapentin and topiramate (Plessas and others 2015) and pregabalin (Sanchis-Mora and others 2019, Thoefner and others 2020). For the short-term studies, quality of life was improved following prescription of topiramate and gabapentin, but not after prescription of carprofen alone (Plessas and others 2015). Prescription of pregabalin improved owner-reported pain scores, mechanical hyperalgesia, cold hyperalgesia, cold allodynia and was efficacious for treatment of neuropathic pain (Sanchis-Mora and others 2019) and SM-associated phantom scratching (Thoefner and others 2020). For the long-term outcome study, 75 per cent of medically managed dogs had an acceptable quality of life at the end of the follow-up period; the most common medication prescribed was gabapentin or pregabalin (Plessas and others 2012). From this, one can conclude there is poor evidence that clinical signs of pain will improve with prescription of carprofen alone, there is some evidence of improvement with combination of carprofen and gabapentin or carprofen and topiramate, and reasonable evidence of improvement with pregabalin.

Anecdotally, a positive response to antacids, such as cimetidine or omeprazole, is reported. The principle is that these drugs reduce CSF production thus reducing the driving force contributing to CM-P and SM. However, studies assessing the effect of omeprazole on CSF production by evaluating the albumen quotient (QAlb; ratio between CSF and serum albumin concentration) did not support a CSF-reducing effect (Girod and others 2016), although the validity of QAlb as a surrogate marker for CSF production was later disputed (Girod and others 2019). More importantly, omeprazole may not achieve a therapeutic choroid plexus concentration,

**BOX 1: MRI FEATURES OF CHIARI-LIKE MALFORMATION-ASSOCIATED PAIN**

- Craniofacial hypoplasia with absent or minuscule frontal sinuses and reduction in maxillary height. Junction between nasal and maxilla forming an angle rather than a slope.
  - Simple explanation: facial features of a ‘forehead’ formed by the frontal bone overlying the brain with no or minuscule frontal sinus, together with well-defined or indented stop and a muzzle which is short in height and length (see Fig 1).
- Rostrotentorial overcrowding resulting in rostral flattening of the forebrain, reduction and ventral displacement of the olfactory bulbs and increased height of the cranium, especially in the occipital region (see Fig 1).
  - Simple explanation: the forebrain changes from a rugby to a football shape (see Fig 2). The olfactory bulbs are small and are directed ventrally rather than rostrally.
- Obstruction of cerebrospinal fluid (CSF) channels: reduction in the cranial and spinal subarachnoid space in addition to ventriculomegaly of all ventricles and cisterns, except the cisterna magna which is often reduced.
  - Simple explanation: on T2-weighted (T2W) imaging there is less ‘white’ hyperintense fluid signal in the CSF space around the brain and spinal cord. By contrast, the CSF spaces within the brain are enlarged.
- Shortening of the basicranium especially the prephenoid bone.
  - Simple explanation: a short skull base.
- Small caudal cranial fossa, the supraoccipital bone is flatter and the opisthion (dorsal foramen magnum) is rostral with respect to the occipital crest (see Fig 1).
  - Simple explanation: the back of the skull containing the hindbrain is small.
- Rostrotentorial neuroparenchyma is displaced dorso-caudally reducing the functional caudotentorial space contributing to a hindbrain herniation.
  - Simple explanation: the forebrain is displaced caudally. The space for the hindbrain is compromised rostrally by the forebrain and caudally by the small caudal skull. The cerebellum loses its rounded shape and is pushed out of the foramen magnum.
Companion animals

so the effect, if any, of antacids on CSF production remains unsubstantiated.

Surgical management

There are three recognised surgical options for management of CM-P and SM-S, but none are entirely satisfactory. To date, no published surgical series has provided MRI evidence of sustained collapse of the syringe postoperatively. Documenting that syrinx does not increase in size is not proof of surgical efficacy. If the postoperative MRI reveals a syrinx which is expanding the spinal cord outline then there is active filling. Likewise, surgical efficacy is not proved by stabilisation of clinical signs, as many dogs can be managed successfully on medical management.

BOX 2: MRI FEATURES OF SYRINGOMYELIA-SPECIFIC SIGNS ASSOCIATED WITH CHIARI-LIKE MALFORMATION

- More extreme brachycephaly than seen with Chiari-like malformation-associated pain (CM-P) (see Box 1).
- Change in the conformation of the craniospinal junction (transitional zone between the brain and the spine) because of craniocervical junction (occiput, atlas and axis) malformation. The atlas is closer to the skull with cervical flexure and acute angulation of the odontoid peg resulting in kinking/elevation of the neuroparenchyma (see Fig 2).
  - Simple explanation: there is a concertina-like flexure of the bones and nervous tissue at the junction between the skull and spine because of rostrocaudal shortening.
- Syringomyelia (SM) is a central cavitation of the spinal cord with fluid that has similar characteristics to cerebrospinal fluid (CSF).
  - For assessment of syrinx severity, transverse images of the widest part of the syrinx are obtained.
    Myelopathic signs in Cavalier King Charles spaniels are associated with a syrinx transverse width of 4 mm or more (SM-S) (Fig a).
- Phantom scratching and cervicotorticollis/scoliosis are associated with extension of the syrinx into the superficial dorsal horn of the cervical spinal cord ipsilateral to the phantom scratching side and/or contralateral to the head tilt.
  - Simple explanation: extension of the syrinx to the edges of cervical spinal cord in the two or 10 o’clock position is associated with phantom scratching and scoliosis. Phantom scratching occurs on the same side as the syrinx. In scoliosis, the head twists down on the opposite side to the syrinx (Fig a).
- Fluid signal-void sign within the syrinx cavity indicates pulsatile or turbulent flow and is a sign of an ‘active’ and filling syrinx more likely to expand (Fig a).
  - Simple explanation: on T2-weighted imaging ‘dark’ hypointense regions within the ‘white’ hyperintense syringe indicates moving fluid.

Surging fluid in the syrinx expands the cavity

- Not all syringes are clinically significant. A quiescent syrinx is centrally located, elliptical on sagittal images and symmetrical, usually circular, on transverse images and results in little or no change to the outline of the spinal cord.
  - An active and filling syrinx is expansive within the spinal cord and generally has an asymmetrical shape on transverse images.
  - Simple explanation: if the spinal cord outline is expanded by the syrinx then it is actively filling and more likely significant. A central located symmetrical ‘hole’ is less likely to be significant (Fig a).
Adjuvant analgesics

| Drug                  | Dose * | Notes and adverse effects |
|-----------------------|--------|---------------------------|
| Gabapentin            | 10–20 mg/kg BID/TID | First-generation α2δ ligand (inhibits excitatory voltage-dependent calcium channels). Can result in mild sedation and poor coordination, especially when therapy is first started. May increase appetite and increased caloric intake may result in weight gain. Therapy should not stop suddenly (ie, the drug should be withdrawn slowly). Avoid xylitol-containing suspensions |
| Pregabalin            | 5–10 mg/kg BID/TID | Second-generation α2δ ligand (inhibits excitatory voltage-dependent calcium channel). Can result in mild sedation and poor coordination, especially when therapy is first started. May increase appetite and increased caloric intake may result in weight gain. Therapy should not stop suddenly (ie, the drug should be withdrawn slowly) |
| Topiramate            | 10 mg/kg TID | Antiepileptic drug with multiple possible mechanisms of action, including carbonic anhydrase inhibition. Little information available for this drug. It should be avoided or used with caution in patients with hepatic or renal disease. The most common adverse effect is sedation and ataxia, which is more likely with polypharmacy. Gastrointestinal adverse effects including inappetence/anorexia may be seen. Irritability, aggression, chewing of digits and facial rubbing have been reported |
| Amantadine            | 3–5 mg/kg PO SID | N-methyl-d-aspartate (NMDA) antagonist which may reduce nociceptive activation as adjunctive therapy (ie, with another drug). Little information available for this drug. The most common adverse effect is sedation and ataxia, which is more likely with polypharmacy. Agitation or gastrointestinal adverse effects may be seen. It should be avoided or used with caution in patients with glaucoma, hepatic disease, renal disease, congestive heart failure, atopic dermatitis or seizure disorders |
| Memantine             | 0.3–1 mg/kg BID | NMDA antagonist which may reduce nociceptive activation as adjunctive therapy. Very little information for this drug. Its use in dogs has been described in compulsive disorder, but not pain (Schneider and others 2009). Its advantage over amantadine is lower cost and a tablet formulation. Systemic review and meta-analysis in human studies suggests it has the potential to decrease pain, but adverse effects of dizziness are common (Kurian and others 2019). Potential adverse effects in dogs include ataxia, tremor and seizures |
| Amitriptyline         | 0.25–2 mg/kg PO SID/BID | Tricyclic antidepressant blocking re-uptake of serotonin and noradrenaline neurotransmitters. The most common adverse effect is sedation, which is more likely with polypharmacy. Other possible adverse effects include hypotension, syncope, dizziness, sleep disturbance, constipation, diarrhoea, palpitations, hyperventilation. Not advised in animals with seizures or epilepsy. Caution in patients with thyroid disorders, urinary retention, hepatic disorders, keratoconjunctivitis sicca, glaucoma, cardiac rhythm disorders or diabetes |

(possible) CSF-reducing drugs

| Drug                  | Dose * | Notes and adverse effects |
|-----------------------|--------|---------------------------|
| Omeprazole            | 0.5–1.5 mg/kg PO SID/BID | Inhibitor of H+/K+-activated ATPase (in the choroid plexus Na+/K+-ATPase regulates the production of CSF). In laboratory rodents long-term therapy reported to result in changes to the stomach lining. However, this effect has not been reported in dogs. Reported adverse effects include nausea, diarrhoea, constipation and skin rashes |
| Cimetidine            | 5–7 mg/kg PO TID | Histamine H2 receptor antagonists (epithelial cells of the choroid plexus possess histamine H2 receptors). Adverse effects with this antacid are rare even at high doses. Liver and kidney toxicity have been reported. In people, cimetidine has been reported to be associated with headache. Cimetidine may increase the kidney clearance of gabapentin |

*BID Twice a day, CSF Cerebrospinal fluid, PO Per os (orally) PRN Pro re nata, SID Once a day, TID Three times a day.
**Companion animals**

Table 4 continued: Unlicensed drugs used in the medical management of CM-P and SM-S

| Drug                                      | Dose *          | Notes and adverse effects                                                                 |
|-------------------------------------------|-----------------|------------------------------------------------------------------------------------------|
| **(Possible) CSF-reducing drugs continued** |                 |                                                                                          |
| Acetazolamide                              | 4–8 mg/kg SID   | Carbonic anhydrase inhibitor (enzyme involved in CSF secretion). Adverse effects are common especially with long-term use and may include anorexia, gastrointestinal signs, bone marrow depression, metabolic derangement (hypokalaemia, hypochlorae mia, hyponatraemia, hyperglycaemia), hepatic insufficiency, hypersensitivity reactions (eg, rash) and CNS signs (sedation, depression, weakness, excitement). |
| **Corticosteroids**                        |                 |                                                                                          |
| Prednisone/Prednisolone/methylprednisolone | 0.5 mg/kg PO SID| Neuroactive steroids modulate pain sensitivity and reduce neuropathic pain. Possible effect on aquaporin-4 expression (water channels) in spinal cord. An option for severe SM-S-associated weakness or phantom scratching. However, long-term use is not recommended due to adverse effects. Chronic use results in muscle loss, thereby increasing weakness and animals are often lethargic and heat intolerant; signs which are easily confused with CM-P and SM-S. Other possible adverse effects include: vomiting; diarrhoea; increased urination and drinking and when combined with diuretics can result in potassium depletion; increased appetite and increased caloric intake that results in weight gain; skin changes and poor hair growth; delayed wound healing and increased susceptibility to infection. This drug should not be given to patients that are pregnant, have diabetes mellitus or kidney disease. |
| **Cannabinoids**                           |                 |                                                                                          |
| Cannabidiol (CBD oil/hemp extract)         | 2 mg/kg BID#    | Cannabinoids act via cannabinoid receptors and affect the activities of many other receptors, ion channels and enzymes. They inhibit release of neurotransmitters and neuropeptides from presynaptic nerve endings, modulate postsynaptic neuron excitability, activate descending inhibitory pain pathways and reduce neural inflammation. Cannabinoids appear to be well tolerated in dogs. Serum biochemistry may show an increase in alkaline phosphatase, presumed due to liver enzyme induction. |

**Cranio-cervical decompression surgery**

Cranio-cervical decompression surgery (foramen magnum decompression) aims to decompress the craniospinal junction by removing the supraoccipital bone with a C1 rostral dorsal laminectomy. Tissue is removed until the cerebellum vermis is well exposed. In my experience, successful decompression also requires removal of the tough atlanto-occipital ligament and a durotomy. Closure varies between surgeons; I favour marsupialisation of the dura and covering the defect with biocompatible collagen matrix (Rusbridge 2007). Others advocate covering the bony defect with an implant typically of titanium mesh. Surgery is successful in reducing pain in approximately 80 per cent of cases; approximately 45 per cent of cases may still have a satisfactory quality of life two years after surgery, although many still receive long-term medication (Rusbridge 2007). Contrary to the analogous human condition, cranio-cervical decompression does not appear to address the factors leading to SM; the syrinx is generally persistent. Much of the clinical improvement is likely attributable to decompression of CSF pathways; that is, surgery is most useful for CM-P rather than SM-S. For some cases, recurrence of signs occurs, often attributed to fibrous tissue adhesions over the foramen magnum; 25 per cent to as many as 50 per cent of cases can eventually deteriorate (Rusbridge 2007). This can occur as early as two months following surgery. There is no convincing evidence that this is less likely with implanted surgery. Fibrous adhesions develop following blood contamination which is common with all surgery.

**Ventricular to peritoneal shunting**

Ventricular to peritoneal shunting may be an option if the ventricles are significantly expanded, or if there is clinical hydrocephalus. This procedure appears to be more successful in facilitating syrinx collapse. However, shunting procedures have a high complication rate, especially subdural haematoma, infection and shunt blockage.

**Syringo to pleural or subarachnoid shunting**

Syringo to pleural or subarachnoid shunting involves placement of a shunting device directly into the syrinx, allowing fluid to drain into the pleural cavity/subarachnoid space, respectively. Because of the risk of shunt blockage,
subarachnoid adhesions and complications relating to the myelotomy, I only use this technique when other surgeries are inappropriate; for example, in the instance of SM secondary to arachnoid webs (Tauro and Rusbridge 2020).

**Which dogs are surgical candidates?**

Surgery is more clearly indicated and most likely to be considered successful in dogs with CM-P (with or without SM) that have responded incompletely or not at all to medical management. In this instance, a craniocervical decompression is probably the surgery of choice. However, if clinical signs reflect SM (eg, phantom scratching or weakness) then this procedure is less likely to be successful because the syrinx persists. Many cases with phantom scratching can be managed medically. Management of weakness is more challenging, especially as there are no surgical reports that provide MRI evidence of long-term postoperative syrinx collapse.

**Complementary therapy**

Failure to control clinical signs and perceived or actual drug adverse effects drive many owners to seek alternative therapy. Anecdotally, acupuncture has been reported to be a useful adjunctive therapy.

![Fig 5: Treatment algorithm for Chiari-like malformation-associated pain (CM-P) and syringomyelia-specific signs (SM-S). Refer to Table 4 for details. CSF Cerebrospinal fluid subarachnoid adhesions and complications relating to the myelotomy, I only use this technique when other surgeries are inappropriate; for example, in the instance of SM secondary to arachnoid webs (Tauro and Rusbridge 2020).

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Companion animals

for some cases. In others, massage may help alleviate signs. Care should be taken as the response to these treatments varies with each animal and some dogs may be more painful afterwards. Spinal manipulation is not recommended because it may cause pain. Exercise should be encouraged, and excessive weight gain discouraged. Hydrotherapy can be useful for some patients, especially those with weakness or proprioceptive deficits.

Genetic factors and breeding advice

The high prevalence, within closely related populations, suggests that SM is inherited in dogs and studies have shown it to be a complex trait, which can be late onset with a moderately high heritability that likely involves genes involved in embryonically active pro-osteogenic signalling pathways. Since the early 2000s, it has been recommended that dogs of breeds predisposed to CM and SM be MRI screened at least twice in their lifetime.

Breeding recommendations based on SM status and ages were formulated in 2006 (Knowler and others 2011). These guidelines concentrated on removing dogs with early onset SM from the breeding pool while maintaining genetic diversity. Early results from this breeding programme indicated that offspring without SM were more common when the parents were both clear of SM. Conversely, offspring with SM were more likely when both parents had SM. In the UK, MRI screens of potential breeding stock can be undertaken through the BVA/Kennel Club’s canine health scheme. However, this does not assess the whole skull or cranial cervical vertebrae nor does it provide an objective measure of risk of developing CM-P or SM. A machine learning approach is being developed with the ultimate aim of creating a simple artificial intelligence tool which will provide an objective measure of risk of developing CM-P or SM in the future (Siteri and others 2019).

Summary

CM and SM is an inherited disorder with a high morbidity in many brachycephalic toy breeds and crosses. It is a pathology affecting the entire skull and craniofacial joint and is characterised by:
- Shortened basicranium;
- Craniofacial hypoplasia;
- Increased cranial height;
- Rostral displacement of atlas and dens;
- Overgrowth of the craniofacial joint;
- Obstruction of CSF flow through the foramen magnum;
- Development of fluid filled cavities in the central spinal cord.

Although some dogs have no clinical signs, others can present with pain (CM-P) and signs relating to spinal cord damage by the syrinx (SM-S). Surgical and medical treatment options are available, but these have limited success, and from a welfare point of view it would be better to implement a screening and breeding programme that limits the occurrence of this disabling disease.

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**COMPANION ANIMALS**

**SELF-ASSESSMENT: NEW CONSIDERATIONS ABOUT CHIARI-LIKE MALFORMATION, SYRINGOMYELIA AND THEIR MANAGEMENT**

In Practice partners with BMJ OnExamination to host self-assessment quizzes for each clinical article. These can be completed online at inpractice.bmj.com

1. Which statement best describes the morphological changes predisposing syringomyelia in brachycephalic toy dog breed dogs?
   a) A cerebellar hemiation into or through the foramen magnum
   b) Shortening of the skull base
   c) A combination of more extreme brachycephaly and cranio cervical junction morphological changes
   d) Small volume caudal cranial fossa (back of skull)

2. Syringomyelia phantom (fictive) scratching is associated with:
   a) Syringomyelia with transverse width less than 4 mm
   b) Syringomyelia with transverse width more than 4 mm
   c) Extension of syrinx into decussating spinthalamic tracts (nociceptive pathway)
   d) A wide syrinx in the mid-cervical region extending to the edge of the spinal cord in the two or 10 o’clock position (area of the superficial dorsal horn)

3. Syringomyelia-associated phantom scratching is characterised by:
   a) Generalised pruritus with feet nibbling but with no obvious skin lesions
   b) Excessive scratching and rubbing to the head and ear region
   c) Rhythmic scratching action towards the neck triggered by light rubbing to the neck or ear region
   d) All of the above

4. What are the most common signs with Chiari-like malformation-associated pain?
   a) Vocalisation, spinal pain, activity change, head scratching and rubbing
   b) Phantom scratching, head scratching and rubbing and spinal pain
   c) Phantom scratching, scoliosis and progressive paresis
   d) Pain face, abnormal head posture when sleeping, aversion to touch

5. Persistent pain associated with Chiari-malformation is best treated with:
   a) Proton pump inhibitor such as omeprazole
   b) Anti-inflammatory drug such as prednisolone
   c) Adjunct analgesic such as gabapentin
   d) Diuretic such as furosemide

6. Dogs with syringomyelia should be:
   a) Completely exercise restricted – walks on the lead to urinate and defecate only
   b) Controlled exercise – 20-minute lead walks two to three times a day
   c) Controlled exercise – unlimited lead walks but no running, jumping or other strenuous exercise
   d) No restrictions on exercise/allowed to exercise to own limits

7. A breeder asks you for advice on preventing syringomyelia. Which of the following would you recommend?
   a) Not to be concerned if they haven’t seen this in their breeding lines
   b) Screening the puppies by MRI and submitting the images to the BVA/Kennel Club’s canine health scheme
   c) Screening the breeding dogs by MRI and submitting the images to the BVA/Kennel Club’s canine health scheme
   d) Screening the breeding dogs through an unofficial MRI scheme so that the results can be kept secret from other breeders and puppy buyers

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ANSWERS: (1) c, (2) d, (3) c, (4) a, (5) c, (6) d, (7) c

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