canine health schemes

Chiari Malformation/ Syringomyelia Scheme



BVA British Veterinary Association

THE KENNEL CLUB
Making a difference for dogs

The **British Veterinary Association** and **the Kennel Club**
— working together for excellence in canine health

What is CM/SM?

Chiari-like malformation (CM) is characterized by a mismatch in size between the brain (which is too large) and the skull (which is too small). This may block the opening from the skull to the spine and alter the flow of cerebrospinal fluid (CSF) around and within the spinal cord. As a result, fluid-filled cavities called syrinxes can develop within the spinal cord, with the condition being called syringomyelia (*"sear-ing-go-my-eelya"*) (SM). The condition has been shown to be inherited, although the mechanism of inheritance is not yet understood.

What is the aim of the Scheme?

The aim of the Scheme is to reduce or eliminate the incidence of inherited CM and SM in dogs. It requires a magnetic resonance imaging (MRI) examination of the brain and upper neck. Although it is expected that the majority of dogs using the CM/SM Scheme will be pedigree toy breeds, the Scheme is open to all dogs, including crossbreeds.

For more information visit **www.bva.co.uk/chs** and click on Canine Health Schemes.

Examples of the MRI features of Canine CM/SM are shown opposite. Figure 1 shows the brain and upper neck spinal cord from a normal dog. Figure 5 shows CM/SM, with the brain pushed out of the back of the skull (arrow) and a fluid-filled cavity (white with asterix) within the spinal cord (grey). Figures 2, 3 and 4 show intermediate grades of CM/SM.

What are the clinical signs?

Although some dogs with a mild CM/SM can seem completely normal, other dogs experience intense pain and may be unable or unwilling to exercise and may have increased sensitivity to touch especially around the head, neck, shoulders and sternum (breast bone). Severe cases may display a characteristic shoulder, neck and/or head scratching where they 'air-scratch' without making contact with the body. Some dogs show neurological signs such as incoordination and weakness.

What breeds are affected?

CM/SM has been shown to be inherited in the Cavalier King Charles Spaniel and the Griffon Bruxellois and is suspected to be inherited in the King Charles Spaniel. Other breeds reported with the condition include among others Maltese, Yorkshire Terriers, Chihuahuas, Papillons, Pomeranians, Affenpinschers, Havanese, Boston Terriers and Staffordshire Bull Terriers.

Does the Scheme help breeders and the pet owner?

Yes, it allows buyers to verify that the parents of their new puppy have been MRI scanned and at what age. Potential buyers can therefore gain reassurance that they are buying from breeders who are performing all the recommended tests to control CM/SM and are using the suggested breeding protocols. Breeders using the Scheme will be demonstrating the highest standards of testing for CM/SM and are doing their best to promote good health and welfare in the dogs they breed.

The Scheme is designed to work with the Kennel Club's Mate Select programme a revolutionary online tool that will enable both occasional and regular breeders to select the most appropriate proposed mating to maximise the chances of producing healthy puppies whilst having the optimum impact on the breed's genetic diversity. Details are available on **www.thekennelclub.org.uk**

How will I arrange an MRI scan for my dog?

There are a number of veterinary centres which offer MRI scanning facilities and which follow the protocol recommended by the Scheme to ensure an adequate quality of the images. The MR images required are DICOM images (Digital Imaging & Communication in Medicine, as used in hospitals). DICOM images are used for security reasons as it is not possible to separate client details from the images.

All dogs must be permanently identified (by microchip or tattoo) before examination and certification. The identification will be verified by the vet performing the MRI prior to the examination and the identification number will be recorded both on the images and on the certificate issued.

You must have the relevant KC owner registration document/related transfer or change of name certificate with you when you present your dog for a MRI scan, if your dog is registered with the Kennel Club. It is possible to scan dogs that are not KC registered; an alternative appropriate form of identification must be provided (e.g. pet name, identification from your veterinary practice or breed club).

Owners must sign a declaration certifying that the details are correct, and that the results may be published by the Kennel Club and also used for research purposes.

All of the above information must be correct at the time of scanning as it is not possible to change the details on DICOM images retrospectively.

What age must my dog be?

The minimum age of a dog for submission to the CM/SM Scheme is one calendar year. There is no upper age limit. As SM is a progressive disease (i.e. dogs can get worse with age) scanning is best performed at

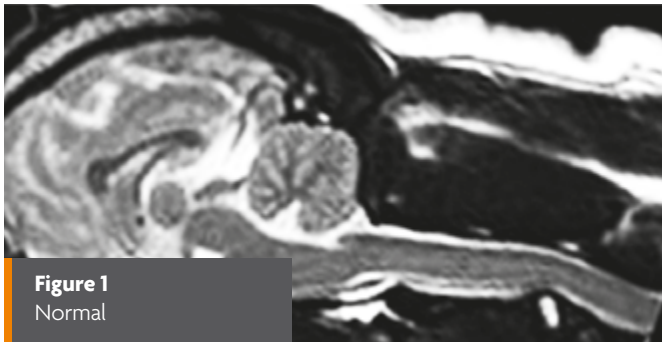


Figure 1
Normal

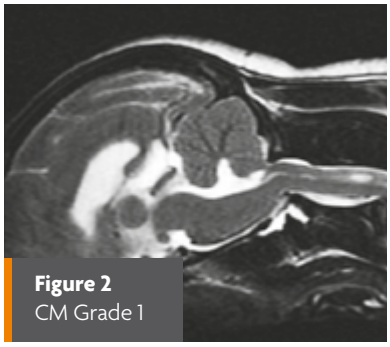


Figure 2
CM Grade 1

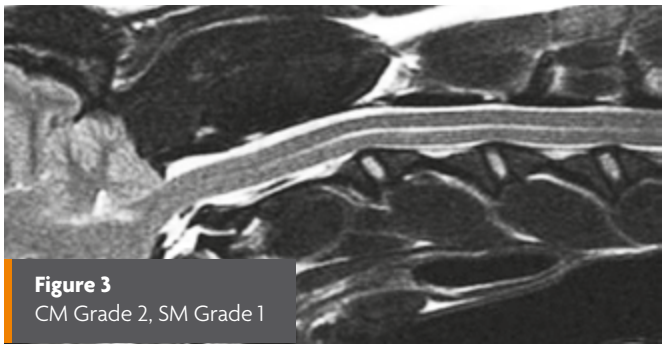


Figure 3
CM Grade 2, SM Grade 1

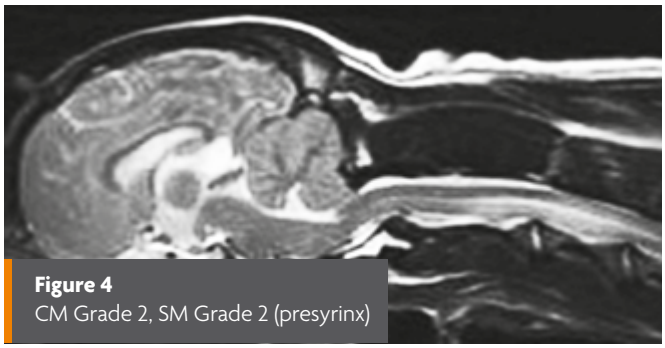


Figure 4
CM Grade 2, SM Grade 2 (presyrinx)

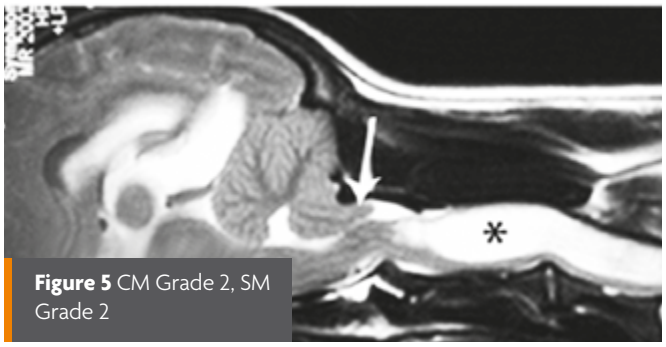


Figure 5 CM Grade 2, SM Grade 2

Chiari-like malformation (CM)

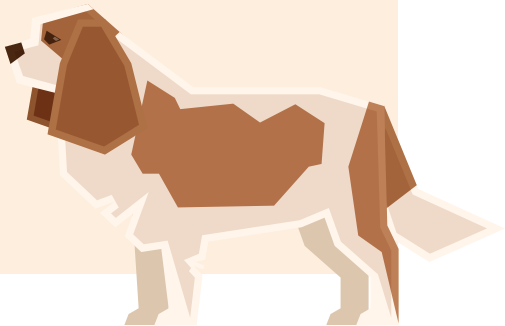
- **Grade 0**—No Chiari malformation
- **Grade 1**—Cerebellum indented (not rounded)
- **Grade 2**—Cerebellum impacted into, or herniated through the opening at the rear of the skull (the foramen magnum).

Syringomyelia (SM)

- **Grade 0**—Normal (no central canal dilation, no presyrinx, no syrinx)
- **Grade 1**—Central canal dilation (CCD) less than 2mm in diameter or a pre-syrinx alone
- **Grade 2**—Syringomyelia (central canal dilation which has an internal diameter of 2mm or greater), or separate syrinx, or pre-syrinx with central canal dilation.

The grade is qualified with a letter indicating the age group at the time of scanning as follows: a = more than five years of age; b = three to five years of age; c = one to three years of age. As SM is a progressive disease the grade is not valid without the qualifying letter.

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five years of age or older, which is late for breeding. Therefore it is recommended that breeders consider scanning their dogs more than once, for example before breeding and again later at five years of age.

Who reviews the MRI scans?

There is a panel of BVA-appointed neurologists and radiologists who will review the MRI scans. The MRI scans are assessed by two scrutineers who will make the final decision. The anticipated turnaround time is about four weeks. There is an appeals procedure.

How are the images graded?

Grading is according to the severity of the CM and SM changes. The age of the dog at the time of scanning is also shown in the grading.

Syringomyelia (SM):

When viewing the images for syringomyelia, a presyrinx, which is cord oedema, is sometimes seen, see Figure 4. This may be transitional and can progress to syringomyelia in time.

What happens to the results?

The results of CM/SM grading will be sent to the submitting veterinary surgeon who will forward the certificate to the owner.

In addition the names and results of KC registered dogs will be sent to the KC for recording on their database and made available via the online Health Tests Results Finder tool on the KC's database. The Kennel Club will forward the information to the Animal Health Trust (AHT) KC Genetics Centre for inclusion in work towards developing Estimated Breeding Values (EBVs) to enhance the reduction of CM/SM in future generations.

Breeding recommendations until relevant EBVs are available:

The aim of these breeding guidelines is to remove dogs with early onset SM from the breeding programme (please note: that in the opinion and experience of the authors, it is believed that due to the complex nature of inheritance of CM/SM it is still possible that affected offspring may arise from parents which are clear from or are only mildly affected by SM. These are recommendations and the BVA and Kennel Club accept no liability for any breeding outcome).

SM grade	Age (years)	Breed to
0a (normal)	Over 5	Any
0b (normal)	3–5	SM grade 0a, 0b, 0c, 1a
0c (normal)	1–3	SM grade 0a, 0b, 1a
1a (CCD)	Over 5	Any
1b (CCD)	3–5	SM grade 0a, 1a
1c (CCD)	1–3	SM grade 0a, 1a
2a (SM)	Over 5	SM grade 0a, 1a
2b (SM)	3–5	SM grade 0a, 1a
2c (SM)	1–3	Do not breed
Any dog with clinical signs of CM/SM	Any	Do not breed

Adapted from: Report from the Chiari-Like Malformation and Syringomyelia Working Group round table. Cappello R, Rusbridge C; Chiari-Like Malformation and Syringomyelia Working Group. *Veterinary Surgery* (2007) 36 509-12.

Note: At the present time, nearly all Cavalier King Charles Spaniels have evidence of CM, so no specific breeding recommendations can be given for CM.

For more information

Visit **www.bva.co.uk** and follow the link to Canine Health Schemes.

Further information about syringomyelia and its management visit **www.veterinary-neurologist.co.uk** and follow the link to syringomyelia.

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For current fees and further information please contact:

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