British Veterinary Association/Kennel Club Chiari malformation / Syringomyelia (CM/SM) Scheme – Procedure Notes

These Procedure Notes are intended to explain the BVA/Kennel Club Chiari malformation / Syringomyelia (CM/SM) Scheme and to provide helpful instruction to those using the Scheme. They are due to be effective from January 2012. They may in future be replaced or modified by further communication.

Introduction

Syringomyelia (SM) is a condition characterised by fluid-filled cavities, called syrinxes, within the spinal cord. In most dogs this occurs in association with a deformity known as Chiari-like malformation (CM). Chiari-like malformation is defined as a mismatch in size between the brain (too big) and the skull (too small) leading to crowding of the cerebellum and brainstem and displacement of the neural structures into the foramen magnum obstructing flow of cerebrospinal fluid (CSF). Both conditions may be demonstrated by magnetic resonance imaging (MRI) scanning. Although SM may be asymptomatic in some dogs it can be associated with neuropathic pain, paresis, ataxia and scoliosis. Syringomyelia has been recognised as a serious problem in the Cavalier King Charles Spaniel (CKCS), Griffon Bruxellois and occasionally in other toy breeds. It has been shown to be a genetically-transmitted condition (Lewis et al., 2012) and in many dogs progresses through the first few years of life (Parker et al., 2011).

Owners participating in the CM/SM Scheme will receive a result regarding their dog's CM and SM status. In addition, in the Cavalier King Charles Spaniel and in some other breeds, results from the CM/SM Scheme may be used to generate Estimated Breeding Values (EBVs) for SM. EBVs are a statistical estimate of the genetic risk for a given disease of individual dogs and a measure of the likelihood of their passing on the disease to their offspring. An EBV is the best method of genetic evaluation available for complex traits and can be calculated for most dogs even if they have not undergone an MRI scan, as long as they are related to dogs which have been scanned. The calculation of EBVs depends on the collection of accurate population-wide data, and for the Scheme to be meaningful and successful it is important that images from EVERY scanned dog be submitted for assessment, whether or not the animal is required for breeding and whatever the state of the spinal cord.

1. The CM/SM Scheme

The main purposes of the CM/SM Scheme are the examination of DICOM digital MR images of the brain and cranial (upper) cervical spinal cord of dogs for changes indicative of CM and SM and the issue of certificates in respect of that examination. If SM is present the certificate will also detail the maximum diameter of the syrinx in the cranial part of the neck. These results and the ensuing EBVs are intended to assist dog breeders in their selection of breeding stock. Breeders wishing to reduce the risk of SM should select potential mates for their breeding stock (both dogs and bitches) based on the EBVs, which will be calculated for KC registered dogs in some breeds. However if the dog is not KC registered or closely related to a KC registered dog then an EBV cannot be generated and owners should refer to the pre-existing breeding guidelines in Appendix 1 and select breeding stock based on the CM/SM result. Owners should also refer to Appendix 1 for breeds and crossbreeds that do not have an EBV scheme.

2. Arranging for MRI and submission to the CM/SM Scheme

The owner or their primary veterinary surgeon should contact an appropriate MRI scanning centre and request that the dog be scanned for assessment under the CM/SM Scheme.

(a) The **minimum** age of a dog for submission to the CM/SM Scheme is one calendar year. There is no upper age limit and it is recommended that breeders determine the MRI status of their breeding stock at three years and again when five years of age. This will provide further information about that individual dog's EBV and therefore the EBV of that individual dog's offspring.

(b) The dog must have permanent identification in the form of a microchip or tattoo. Since microchips interfere with MR images of surrounding soft tissues, breeders should request that microchips are placed no further forwards than the scapulae (shoulder blades).

(c) The following documents must be made available at the time of MRI —

  (i) the Kennel Club (KC) Registration Certificate of the dog, if it is registered with the KC;

  (ii) any related transfer or change of name certificate.

(d) Prior to the MRI scan the owner must complete and sign the first section of the certificate (the Owner's Declaration) verifying that the details given in that section relate to the dog being submitted, that the details are

correct and granting permission for the results to be used in the ways specified. The certificates are provided by the scanning centre and are available to veterinary surgeons from the BVA.

3. Procedure for MRI screening for CM/SM

3.1 Identification of the MR images

The following information MUST be included on the DICOM images at the time of scanning:

(a) EITHER

(i) the Kennel Club registration number (from the top right hand corner of the KC Registration Certificate) for dogs registered with the KC **(no other form of identification for KC registered dogs is acceptable)**

OR

(ii) an appropriate form of identification for dogs *not* registered with the KC (e.g. identification used by the owner's veterinary practice or the breed club);

(b) the dog's microchip or tattoo number, which MUST be confirmed by the veterinary surgeon submitting the MR images and verified on the relevant part of the CM/SM certificate;

(c) the date of MRI.

NB DICOM images which are not correctly identified will be returned to the submitting veterinary surgeon and appropriately identified DICOM images requested. As this information cannot be entered retrospectively onto DICOM images it is essential that it be done correctly at the time of scanning.

3.2 Restraint for MRI scanning

MRI scanning requires that the patient remain motionless (apart from breathing) for several minutes at a time, and therefore general anaesthesia, narcosis or deep sedation is required.

3.3 Images required

The minimum required images are:

(a) Sagittal T1W from the interthalamic adhesion to as far caudally in the neck as possible but at least to C4/C5 intervertebral disc space. The maximum slice thickness permissible is 4mm. The head and neck should be aligned in the sagittal plane and if this is not possible then separate sagittal images of the brain and spinal cord should be obtained so that a midline sagittal image of each is represented. Images will be deemed non-diagnostic if the CNS from the cisterna magna to the C4/C5 intervertebral disc space is not continuously visible in a single sagittal image. The exception to this is if the dog has scoliosis, in which instance dorsal plane images of the relevant area must also be supplied. Dogs with scoliosis as a consequence of SM typically have a marked syringomyelia. If this is not the case the presence of scoliosis may be questioned and the images rejected.

(b) Sagittal T2W as above.

(c) Transverse T1W or T2W images perpendicular to the spinal cord though the maximum width of the syrinx if SM is present, or as a block centred on C3 and extending from at least mid point of the vertebral body of C2 and reaching at least as far caudally as the mid point of the vertebral body of C4. The maximum slice thickness should be 4mm. T2W images are preferred for high field magnets and T1W images are preferred for low field magnets.

Correct positioning is essential to provide MR images of diagnostic quality. The dog's head and neck should be in extension so that the skull base is approximately aligned with the floor of the vertebral canal through C1 and C2. High definition MRI sequences should be selected to ensure optimum visibility of detail.

The images should be checked immediately after acquisition (while the dog is still sedated or anaesthetised) for correct positioning and image quality in case further images must be acquired.

4. Submission procedure

The procedure for submission and grading under the CM/SM Scheme is as follows:

(a) the veterinary surgeon who has performed the MRI scan submits 2 CD copies of the DICOM images together with the part-completed CM/SM certificate, the owner having completed the Owner's Declaration and the veterinary surgeon having signed the Submitting Veterinary Surgeon's Certificate to certify –

(i) the details regarding the submission of the MR images; and

(ii) the microchip or tattoo number once verified.

The CDs may contain scans of more than one dog.

NB The veterinary surgeon who is responsible for the MR scan should check that the breed, sex and colour of the dog correlate with those details in the Owner's Declaration and on the KC Registration Certificate. The veterinary surgeon should also check that the details on the KC Registration Certificate have been accurately and completely transposed by the owner onto the CM/SM certificate i.e. the Kennel Club registered name, number, breed, sex, colour and date of birth.

(b) For payment details see Schedule 1. The BVA charge includes the scrutineers' fees, but **does not** cover the cost of MRI scanning.

(c) The DICOM images are assessed by two scrutineers, from a panel appointed by the BVA. The scrutineers will agree grading for the images, and the completed and signed certificate and the CDs will be returned to the submitting veterinary surgeon; the certificate to be passed on to the owner of the dog.

(d) If the scrutineers are unable to reach a consensus the images will be re-assessed by the Chief Scrutineer, whose decision is final.

If there appear to be any inconsistencies or inaccuracies of completion or identification on the CM/SM certificate the submission will be returned to the veterinary surgeon with relevant comments by the Canine Health Schemes office before the images are examined.

4.1 Rejection of MR images

MR images which are not of acceptable diagnostic quality to the scrutineers will be rejected with constructive comments regarding the reason(s) for rejection, and further images of appropriate quality will be requested. The CDs will be returned; the fee will not be refunded and the certificate will not be completed. When subsequent MR images are submitted for that dog, they must be accompanied by a new certificate and a further fee (see Schedule 1).

5. Grading scheme

The certificate will detail the following:

(a) **Chiari-like malformation (CM)**, graded as follows:

(i) **Grade 0, no CM** – the cerebellum has a rounded shape with signal consistent with cerebrospinal fluid (CSF) between the caudal cerebellar vermis and the foramen magnum.

(ii) **Grade 1 CM** – the cerebellum does not have a rounded shape, i.e. there is indentation by the supraoccipital bone, but there is a signal consistent with CSF between the caudal vermis and the foramen magnum.

(iii) **Grade 2 CM** – the cerebellar vermis is impacted into or herniated through the foramen magnum.

NB At the present time, most Cavalier King Charles Spaniels scanned have Chiari-like malformation.

(b) **Syringomyelia (SM)**, graded as follows:

Syringomyelia (SM) is defined as a fluid-filled cavity that includes or is distinct from the central canal of the spinal cord and is graded according to its maximum internal diameter in a transverse plane.

Pre-syrinx is defined as spinal cord oedema, may be a transitional state prior to development of syringomyelia. Pre-syrinx has the appearance of high signal intensity on T2W images consistent with marked increased fluid content within the spinal cord substance but not of free fluid. On T1W images the spinal cord is either normal or has a slightly hypointense signal.

(i) Grade 0, normal.

(ii) Grade 1, central canal dilation which has a internal diameter of less than 2mm.

(iii) Grade 2, syringomyelia (central canal dilation which has an internal diameter of 2mm or greater), separate syrinx, or pre-syrinx with or without 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. The grade is not valid without the qualifying letter.

6. Results

The results of CM/SM grading are normally sent to the submitting veterinary surgeon within four weeks of receipt by BVA of the correct submission. The arrangements are as follows:

(a) one of the CDs, the completed original certificate (coloured lilac and to be passed to the owner) and a photocopy of the certificate (to be kept by the veterinary surgeon) are returned to the veterinary surgeon who performed the MR scan along with a copy of the Procedure Notes. The other CD is kept by BVA;

(b) the names of Kennel Club registered dogs assessed under the CM/SM Scheme, together with the results, will be sent to the Kennel Club for publication on their website.

Details and results of dogs not registered with the Kennel Club will not be sent to the KC.

6.1 Requests for Results

(a) Pending results:

(i) **an owner must contact the submitting veterinary surgeon, NOT BVA, for results issued under the CM/SM Scheme;**

(ii) a veterinary surgeon may contact BVA on a client's behalf for results of CM/SM assessment but is asked to allow at least four weeks from the date of correct submission before doing so.

(b) Past results:

(i) requests for results which have previously been published should be directed to the Kennel Club;

(ii) any results which have not been published cannot be divulged by BVA and should be sought directly from the owner(s) of the dog;

(iii) a copy of a certificate may be issued by BVA only to the person(s) identified on the CM/SM certificate as the owner(s) of the dog at the time of scoring. Such requests must be made to BVA in writing and a fee will be charged (see Schedule 1).

7. Appeals Procedure

An owner has a right to appeal with regard to the results of a CM/SM assessment. The procedure shall be as follows:

(a) any application for appeal against the result of a CM/SM assessment must be lodged by the owner with BVA in writing **within 45 days** from the date of dispatch from BVA. An appeal certificate (available to the submitting veterinary surgeon from BVA) must be completed within 30 days in a similar way to the original CM/SM certificate and submitted together with the re-grading fee (see Schedule 1). In the event of an appeal being lodged proof of posting will not constitute proof of receipt by BVA;

(b) the MR images will be re-assessed by the Chief Scrutineer, whose decision is final, and the new grading result will be relayed in the normal way (NB the first result may already have been submitted to the KC for publication before the second result is received). In any event, the final result will be sent to the Kennel Club for publication.

SCHEDULE 1

Charges as at 1 January 2012

The scale of fees as at 1st January 2012 is set out below (figures in brackets include VAT). These charges do not include the cost of MR scanning and may be changed from time to time. Any changes will be notified by further communication and will be published on the BVA website at http://www.bva.co.uk/canine_health_schemes/Canine_Health_Schemes.aspx.

BVA charge for scoring per dog	£83.33	(£100.00)
Re-assessment under the appeals procedure	£83.33	(£100.00)

NB DICOM images which are judged by the scrutineers as unsuitable cannot be assessed.

BVA will not refund submission fees for rejected DICOM images.

Copy of the CM/SM certificate (can only be issued to the person(s) identified on the original certificate as the owner(s) of the dog at the time of grading)	£25.00	(£30.00)

In addition to cheques and postal orders, payment may also be made by credit/debit card.

SCHEDULE 2

BVA/KC Chiari Malformation / Syringomyelia Scheme: Panel of Scrutineers as at 1 Janaury 2012

The BVA appointed panel of scrutineers detailed below may be changed from time to time. Any changes will be notified by further communication.

Prof M E Herrtage MA BVSc DVSc DVR DVD DSAM DipECVDI Dip ECVIM MRCVS (Chief Scrutineer)
Mrs R Dennis MA VetMB DVR DipECVDI MRCVS
Dr C Rusbridge BVMS PHD DipECVN MRCVS

REFERENCES

Lewis, T., Rusbridge, C., Knowler, P., Blott, S. & Wooliams, J.A. (2010) Heritability of syringomyelia in Cavalier King Charles spaniels. The Veterinary Journal 183, 345-347

Parker, J.E., Knowler, S.P., Rusbridge, C., Noorman, E.& Jeffery, N.D. (2011) Prevalence of asymptomatic syringomyelia in Cavalier King Charles spaniels. Veterinary Record 168, 667