

Guide to the British Veterinary Association / Kennel Club Chiari malformation / Syringomyelia (CM/SM) Scheme

3. Grading and Breeding Guidelines

How are the images graded?

Grading is according to the severity of the CM and SM changes. 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.

Chiari-like malformation (CM):

Grade 0 - No Chiari malformation

Grade 1 - Cerebellum indented (not rounded)

Grade 2 - Cerebellum impacted into, or herniated through, the foramen magnum.

Syringomyelia (SM):

When viewing the images for syringomyelia, presyrinx, which is cord oedema, is sometimes seen. This may be transitional.

Grade 0 - Normal (no central canal dilation, no presyrinx, no syrinx)

Grade 1 - Central canal dilation (CCD) less than 2mm in diameter

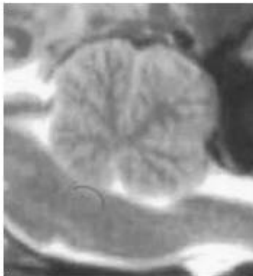
Grade 2 - Syringomyelia (central canal dilation which has an internal diameter of 2mm or greater, separate syrinx, or pre-syrinx with or without central canal dilation).

Why has the age cut off changed from 2.5 to 3 years?

The 2.5 year cut off was based on existing MVD guidelines rather than with scientific justification. A recent publication (Parker et al, 2011) found that the prevalence of SM increased sharply between 0 and 3 years old. After 3 years old the percentage of affected dogs began to plateau. Therefore it was recommended that 3 years be the cut off.

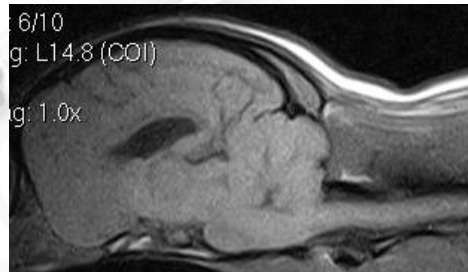
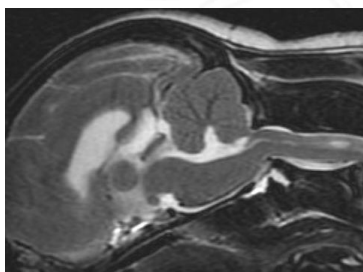
Chiari-like malformation (CM) grading

Grade 0 CM.



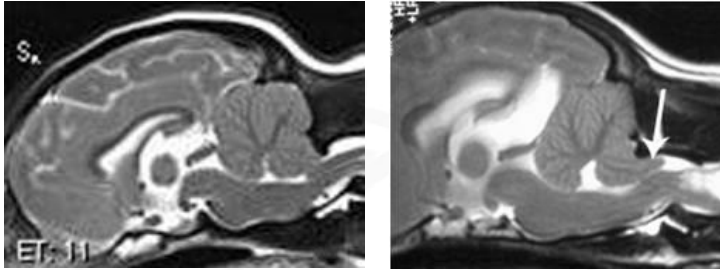
Grade 0 CM. The cerebellum has a rounded shape with signal consistent with cerebrospinal fluid (CSF) between the caudal cerebellar vermis and the foramen magnum. Grade 0 CM or equivalent was reported in 0 of 564 (0%) breeding CKCS MRI submitted in a 24 month period.

Grade 1 CM



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. Grade 1 CM or equivalent (as "mild" CM) was reported in 6 of 564 (1.1%) breeding CKCS MRI submitted in a 24 month period.

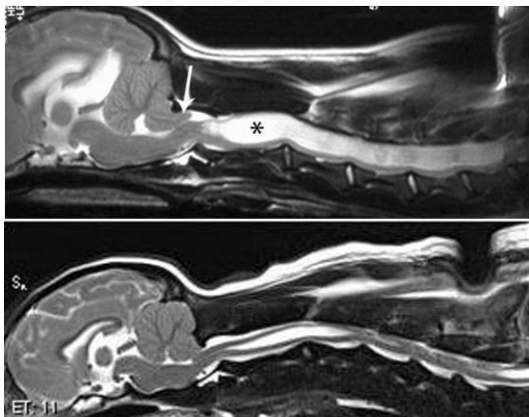
Grade 2 CM



Grade 2 CM the cerebellar vermis is impacted into or herniated through the foramen magnum. Grade 2 CM or equivalent was reported in This was reported (as CM) in 558 of 564 (98.9%) breeding CKCS submitted in a 24 month period.

Why is the size of cerebellar herniation not reported?

The size of cerebellum herniation is not graded because no correlation with genetic risk of syringomyelia has been demonstrated (yet) although dogs with larger cerebellar herniation may have early onset of syringomyelia.



Left

Top image is from a CKCS aged 8 months

Bottom image from a CKCS aged 8 years.

Syringomyelia (SM) grading

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.

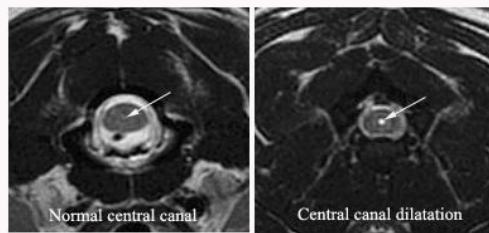


The images above represent a 4mm (left) and 7mm (right) syrinx.

Pre-syrinx is defined as spinal cord oedema and 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. There may be intramedullary widening. The images above were obtained on a low field MRI machine (ESOVATE Vet MRI) and illustrate that although the T2W images are poor quality compared to a high field MRI machine they are still an essential part of a minimum CMSM assessment.

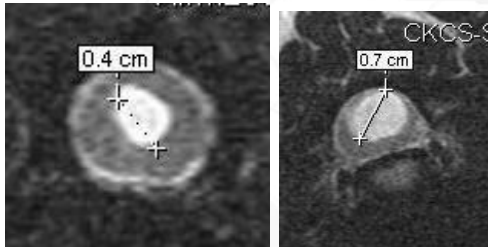
Grade 0 SM – normal , no central canal dilatation

Grade 1 SM -



Central canal dilatation (CCD) less than 2mm in transverse diameter (left)

Grade 2 SM -



central canal dilatation which has an internal diameter of 2mm or greater, separate syrinx, or pre-syrinx with or without central canal dilatation (below)

Breeding guidelines

What if my dog is not KC registered and/or there is no EBV scheme for my breed?

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 breeding guidelines (below) and select breeding stock based on the CM/SM result. Owners should also refer to these breeding guidelines for breeds and crossbreeds that do not have an EBV scheme.

| AGE (years) | SM GRADE | NORMAL | | | CCD | | | SM | | | |
|--|----------|--------------|-----|-----|-----|-----|-----|-----|-----|--------------|--|
| | | 0a | 0b | 0c | 1a | 1b | 1c | 2a | 2b | 2c | |
| >5 | 0a | yes | yes | yes | yes | yes | yes | yes | yes | DO NOT BREED | |
| 3-5 | 0b | yes | yes | yes | yes | | | | | | |
| 1-3 | 0c | yes | yes | | yes | | | | | | |
| >5 | 1a | yes | yes | yes | yes | yes | yes | yes | yes | | |
| 3.5 | 1b | yes | | | yes | | | | | | |
| 1-3 | 1c | yes | | | yes | | | | | | |
| >5 | 2a | yes | | | yes | | | | | | |
| 3-5 | 2b | yes | | | yes | | | | | | |
| 1-3 | 2c | DO NOT BREED | | | | | | | | | |
| Dog with clinical signs CM &/or SM any age | | DO NOT BREED | | | | | | | | | |

The aim of these breeding guidelines is to remove dogs with early onset SM from the breeding programme. Please note: 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 breeding guidelines have been formulated based on the following publications

- Cappello R, Rusbridge C (2007) Report from the Chiari-Like Malformation and Syringomyelia Working Group round table. *Veterinary surgery* : VS 36 (5):509-512. doi:10.1111/j.1532-950X.2007.00298.x
- Knowler SP, McFadyen AK, Rusbridge C (2011) Effectiveness of breeding guidelines for reducing the prevalence of syringomyelia. *The Veterinary record*. doi:10.1136/vr.100062
- Parker JE, Knowler SP, Rusbridge C, Noorman E, Jeffery ND (2011) Prevalence of asymptomatic syringomyelia in Cavalier King Charles spaniels. *The Veterinary record* 168 (25):667. doi:10.1136/vr.d1726

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How do I hear 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 publication on their website. The Kennel Club will forward the information to the Animal Health Trust (AHT) KC Genetics Centre for inclusion in the KC's Mate select programme.