

## Chiari malformation and syringomyelia grading according to the BVA/KC and Utrecht system

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At the moment there are several grading systems. In this document the grading systems of CM and SM are explained as they are reported by us.

### The earliest grading system was the following:

Grade	Age	SM status
A	More than 2,5 years	Absent or a dilation of the central canal of less than 2 mm
C	Less than 2,5 years	Absent or a dilation of the central canal of less than 2 mm
D	More than 2,5 years	Present but no clinical signs
E	Less than 2,5 years	Present but no clinical signs
F	Any age	Present and clinical signs

The BVA/Kennel Club have adapted this system in such a manner that nowadays **the age** and **cerebellum** are also considered.

**There are three age groups:** a = more than five years; b = three to five years; c = one to three years. This is of course vital information as we know that syringomyelia is a late onset disease. Scan result obtained at the age of one may be different when scanned at the age of five.

**Chiari (-like) malformation (CM)**, is, according to the BVA/KC scheme graded as follows:

Grade 0 = no Chiari malformation

Grade 1 = one could say that this is a minimal chiari malformation. Typically there is still some cerebrospinal fluid visible between the caudal cerebellar vermis and the brainstem.

Grade 2 = the cerebellar vermis is completely cramped and pushes out of the foramen magnum.

The age is added hence a grading would be: *CM grade 0c* which means: chiari malformation grade 0 and age group c. We have adapted this slightly in such a fashion that we have added two more groups:

Chiari malformation grading
0 = grade 0
1 = grade 1 as the UK system
2 = grade 2 as the UK system
3 = grade 2 and added to this the herniation and indentation is clearly visible
4 = grade 2 and added to this the herniation and indentation is severe

We also say something about the shape of the cerebellum itself:

Shape
0 = not abnormal
1 = the cerebellum is clover leaf shaped
2 = the cerebellum is flattened at the front side
3 = the cerebellum is flattened at the hind side
4 = Subtentorial herniation is seen
5 = Abnormal shape not listed above

The grading system of the BVA/KC for syringomyelia (SM) is as follows:

Syringomyelia (SM) is defined as a fluid-filled cavity in the spinal cord, whether or not associated with the central canal or the central canal.

A pre - syrinx is defined as spinal edema and can be seen as a precursor to the development of syringomyelia.

SM grade 0 = normal.

SM grade 1 = central channel dilatation where the internal diameter is less than 2 mm.

SM grade 2 = syringomyelia (central channel dilatation which has an internal diameter of at least.

The age group will be added hence the grade would be: *SM grade 0c* if normal and age group c. Hence on each form the BVA/KC grading will be printed on the first page as follows:

**CM grade - age group / SM grade - age group**

We will also provide additional information on the ventricular system and both middle ears (PSOM or middle ear effusion).

Concerning the ventricular system, it will be marked as:

0 = no abnormalities
1 = dilatation of the 4th ventricle
2 = mild dilatation
3 = severe dilatation of the ventricles

Concerning the middle ears:

0 = not visible
1 = only a limited amount of material is visible
2 = there is material visible
3 = the bullae are completely filled with material

Sadly, enough chip artefact does occur. This is caused by the microchip placed in the neck. Low field MRI scanners can produce an artefact and whether this is present or not will be mentioned. Any grading above 1 means that the quality of the scan is of such a quality that the final result could not be estimated and that the current grading could be an underestimate of the real situation. It will be marked as: 0 = no artefact, 1 = present but the MRI scan can be evaluated, 1,5 = MRI scan is evaluated but the result may be less reliable due to the artefact, 2 = cannot be evaluated.

The visibility of the central canal on the sagittal T2 weighted and transverse T1 weighted images will also be mentioned. If a syrinx is visible on a T1 weighted image it will be mentioned. We believe that this may be important as well.

As for the dilation/syrinx we will mention the exact dilation in mm and a special grading. This is similar as measuring it in exact mm's.

<b>Visibility sagittal T2 &amp; transversal T1</b>
0 = not visible
1 = visible sagittal T2
2 = visible S-T2 en TV T1
3 = visible 1-2 mm TV T1
4 = visible 2-3 mm TV T1
5 = visible 3-4 mm TV T1
6 = visible > 4 mm TV T1

The shape of the dilation or syrinx may be important as well and will again be mentioned:

<b>Shape of the central canal or the syrinx</b>
0 = not or circular
1 = presyrinx
2 = CC open to dorsal
3 = CK asymmetrical to aside
4 = separate syrinx

In Belgium they use a different classification and this will be mentioned on the form as well but we advise only to work with the BVA/KC grading and the additional info we provide.

We believe that better phenotyping may help in getting away from CM and SM as published in *Wijnrocx K, Van Bruggen LWL, Eggelmeijer W, Noorman E, Jacques A, Buys N, Janssens S, Mandigers PJJ: Twelve years of chiari-like malformation and syringomyelia scanning in Cavalier King Charles Spaniels in the Netherlands: Towards a more precise phenotype. PloS one 2017, 12(9):e0184893.*

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