



Syringohydromyelia in Cavalier King Charles Spaniels (CKCS) Fact Sheet

Syringohydromyelia is also known as Chiari-type malformation (referring to human disease) and caudal occipital malformation syndrome.

There is a genetic predisposition of Cavalier King Charles Spaniels (CKCS) to develop this neurological abnormality but it can also be seen in other breeds. The diagnosis of Syringohydromyelia is now easily confirmed because MRI is more readily available for veterinary patients.

What is Syringohydromyelia?

Syringohydromyelia on its own is the presence of a cavity within the spinal cord parenchyma filled with cerebrospinal fluid (CSF: the fluid which bathes the brain and the spinal cord), known as a syrinx. The reason why this cavity develops through time in some CKCS is mainly due to an abnormal conformation (shape) of the back of the skull (called the occipital bone).

The caudal part of the brain (cerebellum) doesn't have enough space in the occipital bone and tends to pass through the foramen magnum (exit hole of the spinal cord). This creates a pressure point, giving rise to an abnormal flow of CSF from the brain to the spinal cord. With time this flow disturbance can result in the formation of a syrinx. In some dogs, it can also cause a build up of fluid in the brain called hydrocephalus.

What are the signs of Syringohydromyelia?

Clinical signs can vary widely between dogs and no correlations have been found between the severity of Syringohydromyelia and the severity of the clinical signs.

The most common clinical signs are: twisting of the neck (scoliosis), stiff front limb gait, scratching of the neck and shoulder region, intermittent and unprovoked neck pain, atrophy of the neck muscles, weakness or lameness of the front limbs. They tend to appear with age and their evolution is usually progressive, but can be subacute in some cases.

To complicate the matter, some dogs may present with Syringohydromyelia on MRI without clinical signs. Other neurological diseases may give the same clinical signs.

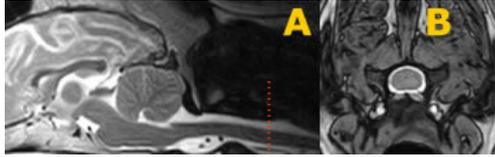
How is Syringohydromyelia diagnosed?

The best way to diagnose Syringohydromyelia is by means of MRI scanning. It is today the best imaging technique to evaluate the spinal cord parenchyma and the back of the brain. The abnormal bone conformation of the skull and the foramen magnum can be seen well with a CT

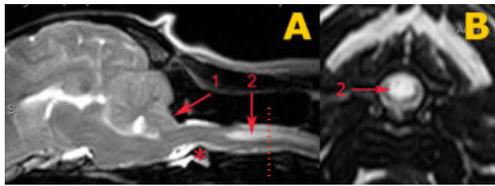
scan (which is better designed to evaluate bone), but the information provided about the cerebellum and the spinal cord is less likely to show up than with MRI. On myelography, Syringohydromyelia usually appears as a swollen cervical spinal cord.

Picture 1 : MRI Image of the the brain (A) and spinal cord (B) of a normal dog. The red interrupted line indicates the level of the image (B).

Picture 2 : Same MRI image of the brain (A) and spinal cord (B) and of an affected CKCS. The cerebellum descent toward the foramen magnum (1), the medulla is kinked (*) and a syrinx is present in the cervical spinal cord (2).



Picture 1



Picture 2

Can you treat Syringohydromyelia ?

Both medical and surgical treatments have been described. At the moment there is no gold standard treatment in dogs although Syringohydromyelia is considered as a surgical disease in man. Depending on the severity of the neurological signs and the age of the patient, a medical treatment is usually recommended in the first instance.

The most common drugs used are prednisolone, acetazolamide and gabapentin. The first two are intended to reduce the CSF production and swelling of the spinal cord. Gabapentin is an anti-epileptic drug used in dogs and humans to treat pain of nervous origin as well as 'pins and needles' sensation. In the absence of a response, a surgical treatment can be proposed.

The aim of the surgical treatment is to restore a normal flow of CSF between the brain and the spinal cord by enlarging the foramen magnum dorsally. Performing the surgery early in the stage of the disease can slow or even stop the evolution of the syrinx.

What is the prognosis for Syringohydromyelia ?

The evolution of Syringohydromyelia is extremely variable between dogs. When clinical signs are present any treatment is more likely to stabilise the clinical signs or slow the course of the disease. As the formation of the syrinx is a slow process, degeneration of the spinal cord can occur at the same time and a complete recovery is unlikely to occur.

It is difficult today to know about the overall prognosis of the surgical treatment, because of the few number of cases treated so far with this method.

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If you are concerned about the health of your pet you should contact your veterinary surgeon.

