Syringohydromyelia
&
The Cavalier King Charles Spaniel

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Syringohydromyelia used to be regarded as a rare condition in veterinary medicine. Now it is one of the more common neurological diseases we see. It has become more common due to the increased availability of MRI and partly due to an increased prevalence in certain breeds most notably the cavalier King Charles spaniel. The CKCS is the 6th top breed in the UK. The breed is gaining popularity in the US where the breed rank is now 32.

Hydromyelia is a dilation of the central canal of the spinal cord. Syringomyelia is a condition where fluid filled cavities develop with the spinal cord tissue. As it is difficult to tell the exact boundaries of the fluid (whether it is just a dilated canal or if the fluid is starting to encroach on the spinal cord tissue) a combined term of hydrosyringomyelia is used.

Ninety percent of syringohydromyelia in people is associated with Chiari malformations. The remaining 10% are associated with other malformations such as intra-arachnoid cysts and Dandy-Walker syndrome. Chiari type I malformation in people is a condition where the cerebellar tonsils and inner part of the cerebellum are elongated into cone-like projections and protrude into the spinal canal.

In dogs there is an analogous condition, characterized by reduced volume in the caudal fossa and caudal displacement of the cerebellar vermis into or through the foramen magnum. Dogs do not have cerebellar tonsils which creates an inconsistency with the historical human condition. A more correct anatomical description for this condition in the dog may be occipital hypoplasia/syringohydromyelia or caudal occipital malformation syndrome (COMS).

Syringohydromyelia is secondary to abnormal cerebrospinal fluid (CSF) dynamics. The exact pathophysiology is unknown. Many theories have been proposed; the water-hammer theory, the suck effect theory, the piston theory, the slosh effect, and the vascular theory. The main debates appear to be 1) does the syrinx form because of increased pressure in the subarachnoid space or because of increased pressure within the spinal cord? 2) What is the source of the fluid within the syrinx- CSF or extracellular? Most researchers think that syringohydromyelia is extracellular fluid, but they do not understand the exact mechanism of how it accumulates.

A few articles regarding hydrosyringomyelia and cavalier King Charles spaniels have been published over the last 5 years. Hereditary is more likely to be autosomal recessive

as both dam and sire must be inbred descendants from certain lines. It is also felt that it is more likely to be of variable penetrance or oligogenic than simple.

There is a broad age range for presentation of clinical signs. Six months to 3 years is the most common age. The onset could be acute or chronic in nature and the progression of signs is variable. One researcher has found that the clinical signs are more severe when the onset of clinical signs occur when the dogs are less than 2 years of age.

The most important clinical sign is pain. Other clinical signs include involuntary scratching at the neck/shoulder region, scoliosis/torticollis, proprioceptive deficits, thoracic limb weakness, ataxia, cerebellar signs, and vestibular signs. Any of the clinical signs may worsen with excitement, weather changes, time of day or activity levels. Some dogs may be overly sensitive to touch on one side of the head, ear, neck, or shoulder.

Humans with syringomyelia report headache, neck pain, back pain, trigeminal pain (facial pain), and radicular pain. Dysesthesia is the most common disabling pain and is described as a burning pain, hyperesthesia, pins and needles, and pressure on the skin.

Diagnosis of syringohydromyelia is made with magnetic resonance imaging (MRI). In human medicine phase-contrast MRI (cine MRI) is used to demonstrate CSF flow. Cine MRI is most useful in cases of borderline Chiari malformations; in demonstrating other CSF blockages; or when the question of when the question of whether or not decompression is needed is not readily answered using traditional MRI.

Dr. March has successfully performed cine MRI in 30 dogs. The affected dogs did have CSF flow abnormalities and neural tissue shifts that corresponded with contractions of the heart. Unfortunately, advanced software and EKG capabilities are needed to do perform this. Myelography primarily shows the outline of the spinal cord. Many dogs with syringohydromyelia may have normal appearing myelograms. The spinal fluid is generally normal or may be mildly inflammatory.

Treatment of this condition consists of either medical therapy or surgical decompression. Analgesics, corticosteroids, and drugs that reduce the CSF production can be used. Mild cases of pain can be controlled by non steroidal anti-inflammatory drugs. Oral opioids or steroids can also be used for control of pain. Gabapentin (anti-convulsant) can be used for severe pain. It has a neuromodulatory effect on the hyperexcitable damaged nervous system which can reduce severe nerve pain.

Proton pump inhibitors and carbonic anhydrase inhibitors can reduce the production of spinal fluid which should decrease the fluid in the syringes. Prednisone reduces both pain and neurological deficits but the mechanism is unknown. Once prednisone has been started it is usually used as a long-term medication. The owners need to be aware of the chronic side effects and should try and use the lowest possible dose.

Surgical intervention is recommended for dogs with significant pain or with worsening neurological signs. The goal is to restore CSF dynamics. The most common surgery is

suboccipital decompression where the hypoplastic occipital bone and the lamina of C1 are surgically removed. The dura can be incised also. The reported success in the small case series varies from no improvement to post operative resolution of the syrinx. Shunting from the syrinx to the subarachnoid space has also been reported.

Dr. Dewey published a recent abstract describing the outcome of 16 dogs undergoing decompressive surgery. 7/16 dogs had resolution of clinical signs and 6/16 dogs had improvement in clinical signs. 3/16 dogs did not improve or were euthanized. Repeat surgery was needed secondary to scar formation in 4/16 dogs.