Papillon Club of America
Health & Genetics

SYRINGOMYELLA & CHIARI MALFORMATION

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and maintained by the Cavalier King Charles Spaniel Club
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Syringomyelia

is a condition whereby fluid cavities develop within the spinal cord. Some refer to syringomyelia as "neck scratcher's disease" because scratching in the air near the neck is a common sign.

Chiari Malformation

is where the back of the skull (posterior fossa) is too small resulting in a gap between the skull and the first cervical vertebrae (atlas) in which the cerebellum is herniated out.

What causes it ?

Syringomyelia is a result of on obstruction to cerebrospinal fluid (CSF) flow. In the normal mammal, the CSF around the brain flows back and forth with the arterial pulse. If this rapid efflux & influx is obstructed then the pressure wave is transmitted down the spinal cord distending it below the blockage. This results in the formation of a cavity or **Syrinx**. Syringomyelia can occur from any blockage in the subarachnoid space (space containing CSF around the brain & spinal cord). However, the most common cause is Chiari Malformation. This condition occurs in many small breeds but is common in the cavalier King Charles Spaniel (conservative estimates at least 50% of the breed although only a proportion are severe enough to have clinical signs).

What are the clinical signs of syringomyelia ?

Pain is the most prominent sign of syringomyelia. This is most commonly localized to the neck region but may be intermittent or difficult to define. Owners often report that their dog isworse at night: when first getting up; during hot or cold temperature extremes; when excited; or related to posture e.g. preferring to sleep with head elevated. They may seem to be overly sensitive to touch on one side of the neck/ear/shoulder/sternum. In addition, some affected dogs scratch at one area of the neck/ear/shoulder/sternum. This is typically one side only, while the dog is moving and sometimes without making skin contact. Some severe cases may have other neurological deficits such as fore and hind limb weakness and ataxia (wobbliness). Facial nerve paralysis, deafness and seizures have also been associated with the condition but a link has yet to be proven.

What age of dog is affected ?

Clinical signs of syringomyelia secondary to chiari malformation are usually recognized between 6 months and 3 years of age. However, dogs of any age may be presented and dogs with more severe disease tend to be presented after 2 years of age.

Do the signs get worse ?

Progression of the disease is variable. Some dogs have the tendency to scratch with mild pain only and have other neurological signs, such as paresis, never or very slowly develop. Others can be severely disabled by pain and neurological deficits within 6 months of the first signs developing. A small syringomyelia may also be found as an incidental finding, with no recognized clinical signs, in the investigation of another neurological disease.

Are there any diseases with similar signs to syringomyelia ?

The main diseases to rule out are other causes of neck pain e.g. disc disease, CNS inflammatory diseases, and other malformations. If scratching or face rubbing is the main sign then skin disease should be eliminated.

How do I know if my dog has Syringomyelia ?

The only way to confirm a diagnosis is by MRI (Magnetic Resonance Imaging). This is essentially a picture of the water content of the body represented in a series of slices (like a loaf of bread). Nervous tissue, which contains a lot of water, is not imaged by x-rays but is shown in great detail by MRI. The syringomyelia can be easily visualized as a pocket of fluid within the spinal cord. In severe cases the syrinx is so wide that only a thin rim of spinal cord remains.

If my dog has been diagnosed with Syringomyelia, what are my options ?

You must decide what is best for you and your pet. Medical management and surgical management are the two primary options.

Medical Management :

Long-term studies of medical management of syringomyelia are not available yet. The drugs used to treat syringomyelia can be divided into 3 types :

1. Analgesics
2. Drugs which reduce CSF production
3. Corticosteroid

Analgesics :

Pain in mild cases may be controlled by non-steroidal anti-inflammatory drugs (NSAIDs) like Rimadyl®, Metacam®, and Previcox®. In more severe cases anticonvulsants, which have a neuromodulatory effect on hyperexcitable damaged nervous system, may be helpful such as Gabapentin (Neurontin®). Oral opioids such as Tramadol, pethidine or methadone are also used.

Corticosteroids :

Corticosteroids are very effective in reducing both pain and neurologic deficits although the exact mechanism is not known. It has been suggested that these drugs reduce CSF pressure however laboratory evidence of this is lacking. Although corticosteroids may be effective in limiting the signs and progression, most dogs require continuous therapy and subsequently develop the side effects of immunosupression, weight gain and skin changes. If there is no alternative, then the lowest possible dose that control signs is used. Alternate day therapy is preferred.

Drugs which reduce CSF production :

Proton pump inhibitors such as omeprazole (Prilosec® or Losec®) can inhibit CSF formation and therefore may be valuable. This drug is unlikely to be useful in the long term as therapy longer than 8 weeks duration is not recommended as this may increase the risk of stomach cancer. Carbonic anhydrase inhibitors such as acetazolamide (Diamox®) also decrease CSF flow and may be helpful in treating syringomyelia although adverse effects of abdominal pain, lethargy, and weakness mat limit long term use. Furosemide (Lasix®) also decreases intracranial pressure and therefore could be useful in the treatment of syringomyelia.

Surgical management :

Surgical management is indicated for dogs with significant pain or with worsening neurological signs. The aim is to restore CSF dynamics and if this can be achieved then the syrinx can resolve. The most common procedure for Chiari malformation is *Suboccipital decompression* where part of the occipital bone and sometimes the cranial dorsal laminae of the atlas are removed to decompress the foramen magnum. The success reported varies from no improvement to post operative resolution of the syrinx. Syringosubarachnoid shunting is sometimes used and id usually successful at significantly reducing the pain but some dogs will still show signs of discomfort/scratching. Also, signs may recur in some dogs after several months to years.

When to have surgery ?

There is more chance of success if the surgery is done early in the course of the disease before permanent damage has occurred. Surgical management is indicated for dogs with significant pain or with worsening neurological signs.

What are the risks of surgery ?

There are major blood vessels in the area and if traumatized, the dog could quickly bleed to death. Although not actually operating on the brain/spinal cord, it is in close proximity and there is a risk of permanent neurological injury. In reality complications from surgery seem to be rare.

Can the disease recur ?

Signs may recur in a proportion of dogs after several months or years due to redevelopment of syringomyelia. The newly created "space" from surgery may fill with scar tissue. If this happens, repeat surgery may be indicated but some owners prefer to continue with medical management (NSAIDs, gabapentin, or corticosteroids).

What post surgery drug treatment would you advise ?

Pets are hospitalized post-op until comfortable enough for morphine-like drugs to be discontinued. They are then discharged on a combination of NSAIDs and Gabapentin. This is withdrawn when the dog is comfortable (about 2-3 weeks).

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