

## **SYRINGOMYELIA**

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### **What is Syringomyelia?**

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

### **What causes it?**

Syringomyelia is a consequence of an obstruction to cerebrospinal fluid (CSF) flow. In the normal mammal, the CSF around the brain shunts back and forth with the arterial pulse. If this rapid efflux and 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 and spinal cord). However, the most common cause is the cerebellum within the foramen magnum (i.e. the back of the brain poking through the hole at the back of the skull). The cerebellum is pushed (herniated) out the skull because there is not enough space since the volume of the back of the skull (occipital bone) is too small. This condition occurs in many small breeds but is common in the cavalier King Charles spaniel (CKCS) (conservative estimates at least 50% of the breed although only a proportion are severe enough to have clinical signs). It is similar to the human condition Chiari malformation (some vets refer to it as Arnold Chiari syndrome which can be confusing as the original description by Arnold was of syringomyelia associated with spina bifida and this is not the case in the CKCS).

### **What are the clinical signs of syringomyelia?**

By far the most important sign of syringomyelia is pain. This is most commonly localised to the neck region but may be difficult to define or intermittent. Owners often report that their dog is worse at night; when first getting up; during hot or cold temperature extremes; when excited; or related to posture e.g. preferring to sleep with their 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 shoulder, ear, neck or sternum. This is typically one side only, while the dog is moving and sometimes without making skin contact. Some dogs, more commonly younger patients, develop a scoliosis (twisted spine). Some severe cases may have other neurological deficits such as fore and hindlimb 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 occipital hypoplasia 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 before two years of age.

### **Do the signs get worse?**

Progression of the disease is very variable. Some dogs have the tendency to scratch with mild pain only and 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

recognised 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 (uncommon in dogs less than two years of age); 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 presented 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 visualised 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 the options?**

No one can make the decision for you about what is best for your dog.

#### 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:

analgesics;

drugs which reduce CSF production;

corticosteroid

#### Analgesics

Pain in mild cases may be controlled by non steroidal anti-inflammatory drugs (NSAIDs) e.g. Rimadyl and Metacam. In more severe cases anticonvulsants, which have a neuromodulatory effect on hyperexcitable damaged nervous system, may be useful, for example gabapentin (Neurontin Pfizer;- these are not licenced for dogs). Oral opioids, e.g. pethidine or methadone are also an alternative.

#### Drugs which reduce CSF production

Proton pump inhibitors such as omeprazole (Losec or Prilosec) can inhibit cerebrospinal fluid formation and therefore may be valuable; clinical data on their use and effectiveness for SM is currently lacking. 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 for stomach cancer.

Carbonic anhydrase inhibitors such as acetazolamide (Diamox; Lederle laboratories) also decrease CSF flow and may also be helpful in treating syringomyelia although adverse effects of abdominal pain, lethargy and weakness may limit long term use.). Furosemide also decreases intracranial pressure and therefore could be useful in the treatment of syringomyelia.

#### Corticosteroids

Corticosteroids are very effective in reducing both pain and neurological 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. They possibly have a direct effect on pain mediators such as substance P. Although corticosteroids may be effective in limiting the signs and progression, most dogs require continuous therapy and subsequently develop the concomitant side effects of immunosuppression, weight gain and skin changes. If there is no

alternative then the lowest possible dose that can control signs is used. Alternate day therapy is preferred.

### 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 like malformation is suboccipital decompression where the hypoplastic occipital bone and sometimes the cranial dorsal laminae of the atlas are removed (with or without a durotomy) to decompress the foramen magnum. The success reported in the small case series varies from no improvement to post operative resolution of the syrinx. Syringosubarachnoid shunting has also been described. In the author's experience surgery is usually successful at significantly reducing the pain but some dogs may still show signs of discomfort /scratching. Also in the author's experience signs may recur in a proportion of dogs after several months/years. One must weigh the risks and benefits of surgery versus medication versus no intervention. Remember, progressive disease means that no action may enable further deterioration.

### 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 traumatised 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?

In the authors' experience signs may recur in a proportion of dogs after several months/years due to redevelopment of syringomyelia. The newly created "space" from surgery may fill in with scar tissue. If this happens, repeat surgery may be indicated; some owner prefer to continue with medical management e.g. with frusemide, NSAIDs, gabapentin or corticosteroids.

### What post surgery drug treatment would you advise?

Dogs are hospitalised until comfortable enough for morphine-like-drugs to be discontinued and then discharged on a combination of non steroidal anti-inflammatory drugs (e.g. Rimadyl) and gabapentin (Neurontin). This is withdrawn when the dog is comfortable (about 2 weeks in most cases).

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