Syringomyelia

Overview:
Syringomyelia (SM) is rapidly emerging as a severe inherited condition in our Cavaliers. It is a progressive neurological disease that varies in severity. Cavaliers unfortunately are affected by SM in larger numbers to any other breed. It is found in all colors, in all lines, and affects both sexes. Signs are usually noticed in dogs between 6 months and 3 years but it has been diagnosed in Cavaliers up to 10 years old. At present the condition can only be identified by MRI scan or by clinical signs. SM occurs when a Cavalier is born with not enough room in the space in the skull that contains the back of the brain. Damage is caused when fluid (CSF) surrounding the brain is forced through a smaller than normal opening, into the spinal cord. The most common symptom is scratching on, or in the air near, the shoulder when the dog is excited or walking on a lead. However this is not the only symptom and it is not always present. Some refer to SM as “neck scratcher’s disease” because scratching the neck is often a sign of the disease. It is similar to the human condition, Chiari type 1 malformation (or Arnold Chiari in some older texts).

Symptoms:
Many symptoms attributed to SM often have nothing to do with this disease so it is important for a vet to eliminate other possibilities first, including PSOM (Primary Secretory Otitis Media) (Link to Other Health Info), or 'glue ear', which causes similar symptoms and can be found in Cavaliers. Allergies to many things, including diet, can also cause dogs to rub their heads on the floor. Ear infections, ear mites, skin conditions or skin irritants like mites or fleas can cause a dog to scratch obsessively or scratch or shake the head and ears. Some dogs are also yelpers, especially when excited. It's a good idea to eliminate more common possibilities first before exploring whether a dog has Syringomyelia. It is important not to read into a Cavalier’s symptoms when they wipe off their face on the carpet after their meal, seek out cold floors on which to sleep, enjoy a good scratch now and again or simply want to be left alone for a while. These are all normal canine behaviors that should not be confused with SM.

The primary symptoms (usually at least one of these is present) are described as:

- **Excessive Scratching** especially while on the lead, and often 'air scratching' where the dog scratches in mid-air, leading to a 'bunny hop' gait as the dog tries to scratch the air with one leg and walk. Sometimes touching the dog's ears brings on scratching.
- **General Pain** is often first noticed because a dog begins yelping or whining or whimpering for no reason. Pain episodes can disappear then return even after a year or more. In some dogs weather changes such as storms or a cold front seem to bring on episodes.
- **Weakness in Limbs** where some dogs may show a lack of coordination. They may limp slightly. Dogs can start to have difficulty getting on and off couches and beds. A paw or leg might go weak. Some dogs will lick at their paws or legs obsessively, often until raw.

The secondary symptoms are described as:

- **Seeking Cool Areas or Restlessness** where an affected dog will shift constantly rather than sleepcomfortably.
- **Head shaking, lip-licking.** Dogs often will shake their heads and ears, yawn excessively (probably an attempt to clear pressure they feel in their heads), or lick at their lips excessively.
- **Head rubbing**. Some dogs start to rub their head from side to side on the floor as if their heads hurt, doing this excessively (NB: normal dogs will do this with pleasure, often before rolling on the floor). They sometimes 'mush' their face against the floor.
- **Digging or pushing**. Some dogs begin to dig obsessively at carpets or sofas. They may run along the length of a sofa pushing themselves against it. Again, this behavior is normal in many dogs; with SM dogs, the activity is frantic.

- **Nerve damage, stiffness, seizures**. This can affect a dog in many ways, from loss of feeling, hearing, or muscular movement. Some dogs have neurological problems with their eyes. Nerve damage seems to be progressive with this condition though some dogs have little or no visible damage and others have severe damage. Some dogs develop a stiffness in the neck, back and/or limbs. In severe cases the neck may bend to the right or left ('neck scoliosis'), or the whole body may bend into a 'C' shape when the dog runs. The head may tilt permanently to one side or the other. The dog may have head tremors. Some dogs begin to have seizures, in some cases, several a day.

Understandably, such descriptions can be confusing – how much scratching is 'excessive', for example? Some people might turn to their vet with such questions, but many have found their vets were unfamiliar with syringomyelia.

Diagnosis:
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). The syringomyelia can be easily visualized as a pocket of fluid (syrinx) within the spinal cord. There would be two reasons to have an MRI performed on your Cavalier: a) to rule out syringomyelia so that the dog can be treated accordingly and b) when it is determined that the dog must have SM surgery.

Managing with Drugs:
Medical management can help but typically does not resolve the clinical signs. Signs in mild cases may be controlled by non steroidal anti-inflammatory drugs (NSAIDs) e.g. Rimadyl. Corticosteroids are very effective in reducing. Although corticosteroids are effective in limiting the signs most dogs require continuous therapy and subsequently develop the concomitant side effects of immunosuppression, weight gait and skin changes but sometimes there is no alternative and the lowest possible dose should be used to control signs. For a CKCS the typical dose would be 5mg prednisolone or 4mg methylprednisolone daily/on alternate days. Gabapentin (Neurontin; Pfizer) is successful in some dogs. This drug, originally patented as an anticonvulsant, is licenced as a neurogenic analgesic for humans. The canine dose is 10-20mg/kg two/three times daily which for a CKCS typically works out at a dose of 100mg two/three times daily. Gabapentin can also be given in combination with NSAIDs. Side effects are minimal and for this reason Gabapentin is preferred over corticosteroids. Oral opioids are also an alternative for example pethidine tablets at 2-10mg/kg three to four times daily or methadone syrup at 0.1-0.5mg/kg three to four times daily. Acupuncture appears to help some dogs. If the dog has seizures, then these can be controlled with phenobarbitol and potassium bromide.

Surgery:
The only option for severe cases of SM is surgery which entails opening the foramen magnum by removing a portion of the occipital bone and usually part of the first vertebrae (foramen magnum decompression surgery). The aim of surgery is to improve the dog's quality of life and/or to stop or reduce further progression. The surgery may not reverse the damage and most dogs still have a tendency to scratch. There is more chance of success if the surgery is done early in the course of the disease before permanent damage has occurred. In older dogs surgery is advised only if the dog is deteriorating. 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. Dogs are hospitalised for a few days until comfortable 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).

General Management:
Some dogs prefer to sleep or eat with their heads up therefore it may be prudent to elevate feeding and water bowls. Purchasing a lightweight harness may help with the scratching when taking dogs for a walk.

Sources: "Introduction to Syringomyelia" by Dr Clare Rusbridge, BVMS DipECVN MRCVS
and "Syringomelia Symptoms" by Karlin Lillington

View:
CKCSC Health Registry, 5+ Year Clear Heart
CKCSC Open Health Registry

List of Neurologists: http://www.acvim.org/Specialist/Search.aspx
MRI Centers: http://www.mrvets.com/mricenter.htm
Look up any Kind of Health Problem: http://www.vetinfo.com/dogindex.html