



Always consult with a veterinarian that you feel comfortable with before diagnosing or treating any disease on your own. This information is for reference only.

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Syringomyelia

Issue Description

Syringomyelia is currently defined as a condition that results in the development of fluid-containing cavities within the parenchyma of the spinal cord as a consequence of abnormal cerebrospinal fluid movement.

Other Names

Neck-Scratcher's Disease, SM

Causes

When there is a blockage or obstruction to the cerebrospinal fluid flow then a pressure wave will be transmitted down the spinal cord and cause the formation of one or more cavities. The most common cause of obstruction is when the cerebellum at the back of the brain is pushed out of the skull because there is not enough space within the occipital bone – a common phenomenon in small breeds and especially in Cavalier King Charles Spaniels, where it is estimated that at least 50% of the breed are affected. Syringomyelia can be congenital (primary) or acquired from trauma, infection or neoplasia.

Breeds Affected

Syringomyelia is one of the most common spinal cord disorders of toy breed dogs and has been reported in the Cavalier King Charles Spaniel, King Charles Spaniel, Griffon Bruxellois, Yorkshire Terrier, Maltese Terrier, Chihuahua, Miniature Dachshund, Miniature & Toy Poodle, Bichon Frise, Pug, Shih Tzu, Pomeranian, Staffordshire Bull Terrier, Boston Terrier, Pekingese, Miniature Pinscher, and French Bulldog.

Symptoms

The most common clinical signs are pain, scratching at the neck and shoulders and walking difficulty. Not all dogs with syringomyelia will present with clinical signs but most dogs will show symptoms of the disease by three years of age. These include restlessness, increasing reluctance to exercise, difficulty moving up or down furniture and uncoordinated movements in the early stages, followed by an uncontrollable urge to scratch the neck area and shoulders, together with a hypersensitivity of the neck area. As the disease progresses, there might be severe pain around the shoulders, neck and head causing the dog to scream and yelp.

More serious cases result in portions of the spinal cord being destroyed, so that the dog contorts his neck and cannot eat or sleep unless its head is held high. In addition, the legs will become progressively weaker and walking becomes increasingly difficult, with some dogs deteriorating to the point of paralysis. The rate of progression varies between individuals – in some, the pain becomes severely disabling and distressing while in others, the condition can be managed by medical or surgical intervention.

Diagnosis

A vet should be asked to rule out basic causes of scratching or discomfort such as ear mites, fleas, and allergies, and then, primary secretory otitis media (PSOM - glue ear), as well as spinal or limb injuries, before assuming that a dog has SM. PSOM can present similar symptoms but is much easier and cheaper to treat. Episodic Falling Syndrome can also present similar symptoms. An MRI scan is normally done to confirm diagnosis of SM (and also will reveal PSOM).

Because of the prevalence, SM is increasingly being considered as important a health issue as mitral valve disease (MVD). Just as many breeders follow the MVD breeding protocol, many breeders are now starting to follow breeding guidelines recommended by international researchers (November 2006), to try to decrease the incidence and severity

of SM in the breed. The guidelines stipulate that breeding dogs be MRI screened (again, unfortunately, the test is very expensive and not widely available yet) and graded according to whether they show the malformation, syrinxes, or both. Neurologists give scanned dogs a signed certificate noting its grade. At least one dog in a breeding pair must be graded A (clear of syrinxes). A limited breeding scheme by a group of Dutch breeders has shown so far that, encouragingly, AxA matings are consistently producing A puppies.

Treatment

Medical management

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 signs partly because of the effect on reducing CSF pressure and possibly because of a direct effect on chemicals which mediate pain. Although corticosteroids are effective in limiting the signs 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 use the lowest possible dose 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 licensed as a neurogenic analgesic for humans. Gabapentin, and other anticonvulsants suppress the firing of hyper excitable damaged nervous system. The canine dose is 10-20 mg/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. Sedation may be seen, especially at higher doses, otherwise the side effects are minimal and on this basis the preference is Gabapentin over corticosteroids. The main disadvantage of Gabapentin is that it is expensive and not licensed for dogs. Oral opioids are also an alternative for example pethidine tablets at 2 ñ 10mg/kg three to four times daily or methadone syrup at 0.1 ñ 0.5mg/kg three to four times daily. Acupuncture appears to help some dogs.

Surgical management

SM is a surgical disease and the most appropriate management is to open 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 reduce the pain improving the dog's quality of life and/or to stop or reduce further progression. If neurological damage has already occurred, the surgery may not reverse the damage and most dogs still have a tendency to scratch.

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 measuring the surgery's success, measure from current condition to the expected further condition and what the disease would have progressed to, rather than the current condition only.

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. Surgery is recommended for dogs with signs at less than 5 years old because progressive disease is likely. In older dogs surgery is advised if the dog is deteriorating.

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?

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 owners prefer to continue with medical management e.g. with NSAIDs, Gabapentin or corticosteroids.

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