

Chiari-like Malformation with Syringohydromyelia (CM-SM)

CM-SM is a congenital disease seen most commonly in Cavalier King Charles Spaniels (CKCS). It is similar to an abnormality in children called Chiari Malformation. The back of the skull has a square rather than rounded shape, causing crowding and compression of the back part of the brain (brain stem and cerebellum).

What is CM-SM

CM-SM is a congenital disease seen most commonly in Cavalier King Charles Spaniels (CKCS). It is similar to an abnormality in children called Chiari Malformation. The back of the skull has a square rather than rounded shape, causing crowding and compression of the back part of the brain (brain stem and cerebellum). The crowding causes the cerebellum to move underneath the back part of the skull through an opening called the foramen magnum. This crowding also interferes with flow of cerebrospinal fluid, and fluid can back up into the brain causing hydrocephalus and accumulate in the spinal cord forming a fluid filled region called a syrinx.

What are the symptoms of CM-SM?

The symptoms of CM-SM are variable. The fluid build up in the spinal cord is suspected to cause an abnormal sensation (similar to a “pins and needles” feeling). This irritation can cause dogs to scratch at the neck/shoulder/ear region or side of the body. Animals often scratch at the air without making contact with the skin (called phantom scratching). The scratching pattern may look like they are trying to scratch their ears. CKCS are also prone to ear problems, so the combination of signs can be confusing.

Other common signs of CM-SM include neck pain, difficulty walking or maintaining balance, head tilt, and facial weakness. The symptoms often occur in episodes, with most dogs behaving normally between episodes. The episodes often occur in association with exercise or excitement, which may be due to increased turbulence of fluid with quick movements. Some dogs with CM-SM show no neurologic signs.

How is CM-SM diagnosed?

MRI is used to document the malformation of the back of the skull, compression of the brain, and accumulation of fluid in the spinal cord. Because CKCS get other forms of neurologic disease (i.e. epilepsy, disc disease) and are prone to ear disease, other causes for neurologic symptoms need to be ruled out before attributing the signs to CM-SM (particularly because CM-SM can be asymptomatic in some dogs).

Vienna

The Hope Center
140 Park St, SE
Vienna, VA 22180
p 703.242.6000
f 703.242.8358

Leesburg

The LifeCentre
165 Fort Evans Rd, NE
Leesburg, VA 20176
p 703.771.2100
f 703.777.9968

Winchester

Valley Veterinary Emergency
and Referral Clinic
210 Costello Dr
Winchester, VA 22602
p 540.450.0177
f 540.662.7870

Woodbridge

Woodbridge Animal Hospital
2703 Caton Hill Rd
Woodbridge, VA 22191
p 703.242.6000
f 703.242.8358

Rockville

Metropolitan Emergency
Animal Clinic
12106 Nebel St
Rockville, MD 20852
p 301.816.0177
f 301.816.0179



What is the treatment for CM-SM?

CM-SM can be treated medically or surgically. Medical management involves administering medications to decrease fluid production to try to slow the expansion of the syrinx. The most common drugs include prednisone (which also acts as an anti-inflammatory and pain medication), omeprazole, lasix, and acetazolamide. Gabapentin is often prescribed to help reduce pain and abnormal sensation.

Surgical treatment for CM-SM involves removing the back part of the skull to alleviate brain compression. Surgery is recommended in dogs with severe clinical signs or those not responding to medical management.

What is the prognosis for CM-SM?

The prognosis for CM-SM is variable. Many dogs improve with medical therapy, although most never return to normal. The side effects of the medications (particularly prednisone) can be very frustrating for some owners and dogs. Surgery is successful in some dogs. Approximately 75% improve with surgery while 25% remain unchanged. Clinical signs recur in about 50% of dogs, and surgery does not resolve the syrinx.

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