

Caudal Occipital Malformation Syndrome/Syringomyelia (aka Chiari-like Malformation)

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SUMMARY:

- 1) In this condition there is a malformation of the skull that can lead to both spine and brain problems.
- 2) This has been described in a number of small breed dog, but is most common in the Cavalier King Charles Spaniel.
- 3) There are both medical and surgical treatments for this disease.

Explanation:

In caudal occipital malformation syndrome (COMS), there is a malformation of the back of the skull. This malformation causes there to not be enough room for the back part of the brain inside the skull. Consequently, a part of the brain called the cerebellum is pushed from its normal position and starts to exit the skull (called herniation). This abnormal arrangement causes the spinal fluid in this area to flow abnormally. This abnormal flow can lead to pocket of fluid building up in the spinal cord (called syringomyelia OR SM) as well as an enlargement of the normal fluid spaces in the brain (called ventriculomegaly or hydrocephalus depending on the severity).

This disease is most commonly diagnosed in young adult Cavalier King Charles Spaniels, however it is can be found in any age and has been diagnosed in many different small breed dogs. Some reports say that up to 95% of Cavalier King Charles Spaniels have the malformation, however a much smaller percentage have symptoms.

Symptoms:

- 1) Scratching at the neck and ears. Often dogs will make a scratching motion and not actually make contact with the skin (called “phantom scratching” or “air guitar”).
- 2) Head and Neck Pain
- 3) Incoordination and weakness
- 4) Abnormal curvature of the neck (Scoliosis)
- 5) Seizures, facial paralysis and deafness are sometimes reported.

Diagnostics:

COMS/SM is best diagnosed by MRI of the brain and spinal cord. Other testing, such as CT scans and BAER testing, may also be utilized depending on the availability of MRI in your area.

Treatment:

There are both medical and surgical treatments for COMS/SM.

Medications for this disease are aimed at reducing the amount of spinal fluid and treating symptoms like pain and seizures. These treatments cannot fix the underlying problem though, and therefore medical treatment eventually fails in a significant amount of dogs. Sometimes more severely affected patients will



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not respond at all to medications. However, in less severely affected dogs, the medicines may work temporarily, and sometimes even for years.

At surgery we remove the abnormal piece of bone that is pinching the brain, which allows the spinal fluid to flow more naturally. The area is then covered with a material to help prevent scar formation. Earlier versions of this surgery had a higher complication rate because scar tissue would form at the site of the removed bone and cause compression again. However, by reforming the skull with a plate this complication rate is dramatically reduced. Depending on the exact nature of your pet's malformation, sometimes your neurologist will recommend a different procedure, such as placing a shunt to drain excess fluid from the brain.

We see improvement in approximately 80% of dogs with surgery. Some dogs do require some medication still after surgery, however usually it is a much lower amount of medication and the symptoms are better controlled.

Medications:

1. **Gabapentin:** Gabapentin is used to treat pain and abnormal sensations (like tingling and burning). If this medication is not successful, other medications like pregabalin or amantadine might be tried.
2. **Prednisone or other steroids:** This type of medication helps reduce the production of spinal fluid in the brain and also reduces inflammation.
3. **Omeprazole:** This type of medication helps reduce the production of spinal fluid in the brain and also can help with an GI upset that develops secondary to prednisone.
4. **Pain medication:** Some dogs will require additional pain medication, such as tramadol or other opioids.
5. **Seizure medications:** If seizures are present, seizure medication may also be recommended.

Monitoring:

Please monitor for any change in the symptoms listed above. Notify your veterinarian if symptoms are getting worse or if your pet is experiencing unpleasant side effects from medications. After initial diagnosis, it is recommended to check in frequently with your veterinarian so the correct medication schedule can be determined. Rechecks are recommended at least every 6 months so your veterinarian may monitor your pet's disease and check for any side effects of medications.

Primary Secretory Otitis Media (PSOM):

There is a congenital ear problem found primarily in Cavaliers that can mimic many of the symptoms of COMS/SM, including scratching, as well as head and neck pain. In this disease there is a buildup of mucus without signs of infection deep in the ear. This stretches the eardrum causing pain. Much like COMS/SM, PSOM is also diagnosed on MRI/CT scans. Treatment involves draining the mucus by a variety of methods. It is often mentioned when we are discussing COMS/SM because the symptoms are so similar and without MRI/CT some patients may not be treated for the correct disease.

