Caudal Occipital Malformation Syndrome

Robert L Bergman, DVM, MS, Dip ACVIM
Carolina Veterinary Specialists

With the increasing availability of MRI, as well as improved education of pet owners, caudal occipital malformation syndrome (COMS) is being recognized more frequently. The problem is similar to a Chiari type 1 malformation recognized in humans. With this syndrome, there is crowding of the cerebellum due to compression by the caudal aspect of the skull above the foramen magnum. It has been shown in the Cavalier King Charles spaniel (CKCS) that the brain is too large for the contents of the caudal aspect of the skull (caudal fossa). This may result in herniation of the cerebellum, kinking of the medulla, and changes in the normal fluid dynamics of CSF. It may also lead to abnormal CSF flow in the spinal cord. Syringomyelia (fluid accumulation within spinal cord) and hydromyelia (dilated central canal) are commonly found in dogs with COMS. Syringomyelia is commonly referenced as SM. The formation of a syrinx many times causes compression of the dorsal horn of the spinal cord, resulting in changes in sensory perception. This is the cause for neuropathic pain associated with this disease. It has been shown that certain mediators of inflammation including substance P and IL-6 are up regulated in this portion of the spinal cord, exacerbating the condition. Syrinx formation may be discontinuous and may be present in cervical, thoracic, and lumbar spinal cord segments. Caudal occipital malformation can cause pain due to compression of the brainstem or the first cervical nerve.

There is some disagreement with what constitutes SM. Dilation of the central canal >2 mm is known to be associated with neuropathic pain. However, some have suggested dilation greater than 1 mm constitutes SM. The presence of syringomyelia is usually associated with a caudal occipital malformation, but it is possible to have SM without a malformation. Furthermore, the incidence of asymptomatic SM is up to 70% in older CKCS.

This problem is most prevalent in the Cavalier King Charles spaniel. Extensive study has been done on this breed, and the prevalence of COMs is 95%, while the presence of COMS with syringomyelia is 46%. In Cavalier’s that are asymptomatic for the condition but have changes on MRI, the presence of a syrinx significantly increases the risk for developing clinical signs later in life. However if no clinical signs are evident by age 6, most will remain asymptomatic. The inheritance is complex. A recent study demonstrated that the estimated heritability of symptomatic SM was about 0.81. The Brussels Griffon dog (Griffon Bruxellois) has also been reported to have a high incidence of this condition. In a recent report on this breed, the prevalence of COMs was 65%. Of those dogs, 78% had some degree of SM. Other breeds (small breed dogs) include Yorkshire Terriers and Toy poodles. Clinical signs of this condition include severe neck pain, phantom scratching, scratching the side of the face or shoulder, and ataxia. Pain may be nonspecific and intermittent. Sometimes owners complain of yelping for no apparent reason or when the dog is picked up in a specific manner. Some breeds, notably the Brussels Griffon, have a different presentation of this condition. These dogs more commonly
have weakness and ataxia and less commonly have neck pain. They also do not tend to have significant phantom scratching. Clinical signs of the condition may be present before 1 year of age or may be recognized much later in life. Another condition that occurs frequently in the KCKS is primary secretory otitis media (PSOMs or “glue ear”). Clinical signs of this condition include head and neck pain, scratching of the ears, facial paralysis, head tilt, and hearing loss. This condition may occur with SM or may be a confounding factor.

The problem is diagnosed by MRI of the brain and cervical spinal cord. A mid-sagittal T2 view of the brain and upper cervical spinal cord are the most helpful views. Positioning of the head for MRI is important for an accurate assessment of this condition.

Treatment of this condition is controversial and specific guidelines have yet to be determined. Many dogs with this condition are asymptomatic and most likely to not warrant treatment. Furthermore, some dogs have coexisting conditions including GME, IVDD, PSOMs, or AA instability. The treatment of CM/SM consists of medical management and surgery. Medical management usually involves the use of multiple drugs. Analgesic drugs modulate noxious stimuli and pain. Gabapentin (10 mg/kg PO q 8 hours) modulates pain in the dorsal horn of the spinal cord and can be helpful for controlling pain and scratching. Side effects include sedation and ataxia. Pregabalin (2-4 mg/kg PO q 12 hours) is a newer analogue of gabapentin. Some consider this to be more effective than gabapentin. It is more expensive and is a controlled drug. Opiate drugs may also be used. Tramadol (2-4 mg/kg PO q 8-12 hours) is helpful in combination with Pregabalin or Gabapentin. Other types of drugs used for neuropathic pain include antidepressants (amitriptyline) and NMDA antagonists (amantadine). Drugs that reduce cerebrospinal fluid production can also be used. Omeprazole, a proton pump inhibitor, has been shown to reduce CSF production 26% experimentally when given intrathecally. Suggested dosing is 10 mg for dogs less than 20 kg and 20 mg for dogs larger than 20 kg PO q 24 hours. Diarrhea is the most common side effect. Other drugs that reduce CSF production include corticosteroids, acetazolamide, and furosemide. Corticosteroids are anti-inflammatory drugs that reduce CSF production and reduce expression of substance P in the spinal cord. The recommended dose is 1 mg/kg/day, and it is recommended that this be tapered to alternate day therapy if possible.

Surgery is advocated by some as the preferred method of treatment in symptomatic dogs. However, others recommend surgery in severe cases that do not respond to medical treatment. Surgery consists of enlarging the foramen magnum (foramen magnum decompression). The prognosis is variable, and some dogs will develop a recurrence of clinical signs if extensive epidural fibrosis develops within the syrinx, thought to be due to prolonged pressure. The presence of an asymmetrical and dorsally located syrinx is associated with a poorer outcome following surgery. This may be due to spinothalamic tract involvement in the dorsal horn of the spinal cord through connections of the superficial laminae. Surgery is considered successful in the short term about 80% of the time with foramen magnum decompression. However, this improvement may be temporary if fibrosis occurs at the surgery site, thus recreating the original compression. As mentioned, size and location of the syrinx also have a role in outcome. A variation of foramen magnum decompression is the addition of a protective plate to cover
the skull defect. This technique known as cranioplasty is intended to reduce potential compression from fibrosis over the surgery site. Dewey and others have reported a recurrence rate of 1% when cranioplasty is used. Following surgery, pain management is continued. Owners are educated regarding expectations for improvement. In general, improvement is a gradual process, once the initial pain from surgery resolves. Improvement can be variable and may take a few days or a few months. Scratching will persist in many instances due to the presence of the syrinx.
Suggested Reading:


**Atlantoaxial Subluxation**

Atlantoaxial (AA) subluxation may be congenital or secondary to trauma. This problem tends to occur in young toy or miniature breed dogs, however it may be identified in older or larger dogs. Commonly affected breeds include Yorkshire terriers, miniature poodles, Japanese Chin, and Chihuahuas. There may be rupture of the transverse atlantal ligament allowing the axis to rotate dorsally. There is compression of the spinal cord between the axis and atlas due to dorsal displacement of the axis into the vertebral canal. It may be complicated by the dens, if the dens is present. This can be a chronic or an acute event.

Clinical signs of this disease are variable and are often misinterpreted as intervertebral disk disease in a young dog. The animal may present with neck pain, severe ataxia, or even tetraplegia. Generally, these signs are noticed early in life but can occur later. Signs may progress and may be associated with trauma. It is becoming increasingly apparent that this is not necessarily one distinct disease. Caudal occipital malformation-syringomyelia or occipital overlap syndrome may be comorbidities. Furthermore, in some dogs, the finding of AA instability is incidental, and in fact the dog has an inflammatory disease (GME). If atlantoaxial instability is suspected, be very careful when manipulating the neck. Neurological deficits should localize to C1-5.

The diagnosis of this disease begins with a lateral radiograph. Cervical radiographs are best done with the animal awake because general anesthesia may allow the neck to move excessively. There may be significant separation between the dorsal aspect of C1 and C2. This can be variable and may be difficult to definitively diagnose. Careful flexion and extension with survey radiographs or with fluoroscopy may identify instability. The ventrodorsal views of C1 and C2 should be evaluated for the presence of a dens. Ideally, MRI and CSF analysis should be done to rule out other diseases. However, this is not always practical due to cost. The owner should be made aware of the possibility of other diseases.

The treatment of this disease involves surgical stabilization/fusion of C1-2. There are significant risks associated with this surgery including death and implant failure. However, most of these dogs do well, even after severe initial signs in acute cases. If surgery is not an option, a neck brace may be used for at least 4-6 weeks. Unfortunately, this may not result in successful fusion. The client should be warned that there is the possibility for the development of severe signs, even possible death, if the animal becomes more unstable.

There have been multiple methods described for surgical fusion of C1-2 and include both dorsal and ventral approaches. In our practice, we use a ventral approach to this site. If the dens has been fractured (Hangman’s fracture), this may also need to be removed.
Suggested Reading:
