CAUDAL OCCIPITAL MALFORMATION SYNDROME IN DOGS

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INTRODUCTION
Caudal occipital malformation syndrome (COMS) is the canine analog of human Chiari type I malformation. The disorder is a congenital anomaly of the caudal occipital region of the skull that leads to overcrowding of the caudal fossa and compression of the cervicomedullary junction at the level of the foramen magnum. Both direct bony compression and progressive meningeal hypertrophy in the region of the dorsal cervicomedullary junction are thought to lead to abnormalities of cerebrospinal fluid (CSF) flow dynamics. These CSF flow changes often lead to fluid accumulation within the spinal cord (i.e., syringohydromyelia). Clinical signs of central nervous system (CNS) dysfunction referable to the brain and/or spinal cord can result from COMS.

PATHOPHYSIOLOGY OF COMS
Although the cause of COMS is unknown, it is suspected to be a genetically transmitted developmental disorder of the occipital bone mesoderm. In patients with COMS, there tends to be some level of cerebellar compression, as well as constriction of the cervicomedullary junction in the vicinity of the foramen magnum. With chronic bony compression at the cervicomedullary junction, and probable turbulent CSF flow and pressure changes in this region, it is thought that the underlying meninges become hypertrophied with time. Such hypertrophy has been documented in both humans with Chiari type I malformation and dogs with COMS. There are numerous theories to explain the development and propagation of syringohydromyelia cavities in patients with COMS. An in-depth discussion of these theories (e.g., “water-hammer” effect, “suck”, “slosh”, “ball-valve” effect) is beyond the scope of this presentation. Common to all of these theories is obstruction of normal CSF flow at the level of the cervicomedullary junction.

MRI CHARACTERISTICS OF COMS
Imaging modalities other than MRI are not likely to consistently diagnose COMS. Magnetic resonance imaging is also the preferred method for diagnosing syringohydromyelia. Characteristic MR imaging features of COMS patients include the following: attenuation/obliteration of the dorsal subarachnoid space at the cervicomedullary junction; rostral displacement of the caudal cerebellum by the occiput; cervical syringohydromyelia; cerebellar herniation through the foramen magnum, and: “kinked” appearance of the caudal medulla. The most critical image to evaluate is a mid-sagittal T2-weighted view.

CLINICAL FEATURES OF COMS
Caudal occipital malformation syndrome appears to be confined to small-breed dogs. The most commonly reported breed affected by COMS is the cavalier King Charles spaniel. Other breeds reported include Yorkshire terrier, miniature/toy Poodle, Maltese, Pomeranian, Pug, Chihuahua, Bichon Frise, miniature Pinscher, West Highland White terrier, Shih Tzu, Pekingese, and French Bulldog. The mean age at presentation is between 4 and 6 years of age, although there is a wide age range. Clinical manifestations of COMS include multifocal CNS dysfunction, cervical myelopathy, cerebellovestibular dysfunction, and forebrain dysfunction. Occasionally, a dog with syringohydromyelia will demonstrate torticollis. A unique clinical sign of COMS, presumably related to cervical syringohydromyelia, is persistent scratching at the shoulder, neck, and head regions. Cerebrospinal fluid results are typically normal with COMS; occasionally, there is a mild mononuclear pleocytosis and/or a slight elevation in protein level.
TREATMENT AND PROGNOSIS OF COMS

The preferred treatment for people with symptomatic Chiari type I malformation is foramen magnum decompression (FMD). Treatment of COMS patients with oral glucocorticoids (e.g., prednisone-0.5 mg/kg, BID) often results in clinical improvement. The author has also had considerable success with oral gabapentin (10 mg/kg TID) in alleviating scratching behavior. Although case numbers are limited, it appears that approximately half of COMS dogs treated medically will progress after 1-2 years despite therapy. The author and colleagues have recently investigated a FMD procedure for COMS. Sustained resolution or improvement of clinical signs was achieved in approximately 80% of patients. Approximately 25% of these dogs formed exuberant scar tissue in the region of the FMD within several months of surgery and required re-operation (due to compression at the foramen magnum). Adjunctive surgical procedures used with success in people with Chiari type I malformation (e.g., syringopleural/syringoperitoneal shunting) have not yet been evaluated for COMS in dogs.

List of References