Chiari-Like Malformation and Syringomyelia (CM/SM)

Chiari-like malformation (CM) and syringomyelia (SM) often occur together, although both may occur independently of the other. Syringomyelia is a condition characterized by the presence of a fluid-filled cavity (syrinx) or cavities within the parenchyma of the spinal cord. SM is secondary to abnormal cerebrospinal fluid movement and is usually associated with Chiari-like malformation, although it may be associated with other conditions, such as congenital malformations, trauma, inflammation, and neoplasia. Chiari-like malformation is defined as a decreased caudal fossa volume with herniation of the cerebellum and often the brainstem into or through the foramen magnum. In people, this condition is referred to as Chiari malformation, which has several types.

The term syringomyelia is accepted to describe fluid accumulation within the spinal cord, whether it be secondary to central canal dilation (hydromyelia) or secondary to fluid accumulation within the spinal cord parenchyma (syringomyelia or syringohydromyelia). It is difficult to determine the location of the fluid using magnetic resonance imaging (MRI), and these cavities often communicate with each other. Syringomyelia frequently occurs with Chiari-like malformation in dogs and the terms Chiari-like malformation and syringomyelia (CM/SM) have been adopted to describe the canine condition.

Clinical Signs

Onset of signs may be acute or chronic in dogs ranging from six months to ten years of age. The most common sign of CM/SM is pain, predominately isolated to the cervical region, occurring in 35% of affected dogs and 80% of people with the similar condition. Syrinx width, measured by MRI, has been shown to be the strongest predictor of pain in dogs where a wider syrinx was significantly associated with discomfort. Additionally, the location of the syrinx within the dorsal aspect of the spinal cord affecting the dorsal horn is thought to be one mechanism behind the development of pain, specifically neuropathic pain. Neuropathic pain is secondary to disordered processing of sensory information within the nervous system and results in spontaneous pain, paresthesia, dysthesia, allodynia, or hyperpathia. As a result, dogs may dislike touch to the skin of their neck, or they may scratch, with or without making contact to the skin on their neck. This “phantom scratching” has frequently been described in affected dogs. Pain may also be to CM alone, as seen in dogs without SM secondary to compression of the brainstem or first cervical nerve.

Other clinical signs depend on the location of the syrinx, although the cervical spinal cord is predominately affected. These clinical signs include scoliosis and neurological deficits relating to cervical spinal cord dysfunction. Intracranial signs, such as facial paresis and vestibular dysfunction, have also been reported. However, dogs may also be asymptomatic for CM/SM.

Pathogenesis

The human classification of Chiari type I, which is the most similar to the canine condition, necessitates elongation and caudal displacement of the cerebellar tonsils (vermis and paravermal lobes) through the foramen magnum into the cranial cervical vertebral canal. A similar condition has been documented in dogs, particularly toy or small-breed dogs. Predisposition to CM/SM has been seen in Cavalier King Charles Spaniels (CKCS) and Brussels Griffon dogs. In CKCS, CM/SM is a hereditary condition, possibly autosomal recessive with incomplete penetrance. CM is caused by congenital hypoplasia of the supraocципital bone resulting in overcrowding of the structures within the caudal fossa. As a result, the cerebellum herniates through or into the foramen magnum, the medulla becomes kinked, and the dorsal subarachnoid space at the craniocervical junction is obstructed. The flow of CSF through the foramen magnum is disrupted as a result. Many theories behind why syringomyelia develops as a result of this obstruction have been postulated, although a single theory has not been proven.

Diagnosis

These structural abnormalities are best diagnosed with MRI, but they may be clinically silent; therefore, their significance must be carefully considered when such abnormalities are discovered.
Treatment and Prognosis

Treatment may not be necessary in asymptomatic dogs or dogs with mild nonprogressive signs. Dogs exhibiting pain, more severe neurological deficits, or progressive signs can be treated either medically or surgically. Typically, medical therapy is pursued, initially involving the use of analgesics and drugs that reduce CSF formation. Furosemide (1–2 mg/kg orally q 12 hrs) and prednisone (0.5–1 mg/kg orally 24 hrs, tapering dose) are frequently used. Treatment of neuropathic pain with drugs such as Gabapentin (10 mg/kg PO q 8 hrs) is also an important aspect of therapy.

Approximately 70% of patients show some improvement, but it is rarely complete. If medical therapy does not alleviate the clinical signs, surgical decompression of the foramen magnum has been suggested (suboccipital craniectomy) and is the treatment of choice in people. Foramen magnum decompression has been performed in dogs with success rates reported at about 80%; however, recurrence is common and neuropathic pain may persist, requiring continued medical therapy. Additionally, multiple surgeries may be required if scar tissue develops at the surgical site obstructing CSF flow, although cranioplasty may reduce the likelihood of this complication. Improvement may not be a result in the reduction of syrinx size, which usually persists.

Steroid Responsive Meningitis-Arteritis

A severe form of steroid responsive meningitis-arteritis (SRMA) has been reported in beagles, Bernese mountain dogs, boxers, German short-haired pointers, and sporadically in other breeds. This condition has a worldwide distribution and represents one of the most important inflammatory diseases of the canine CNS. Beagles, especially, but not exclusively those in laboratory-bred colonies, appear at risk. In beagles, the condition has been termed beagle pain syndrome, necrotizing vasculitis, polyarteritis, panarteritis, juvenile polyarteritis syndrome, and primary periarteritis. In other breeds, this condition previously appears under the terms necrotizing vasculitis, corticosteroid-responsive meningitis, aseptic supplicative meningitis, and corticosteroid-responsive meningomyelitis. This plethora of terminology reflects not only the dearth of knowledge about this condition but also highlights important clinical signs such as pain, improvement following corticosteroid medication, and histologic involvement of the meninges and blood vessels.

Affected animals usually are most commonly young adults between eight and eighteen months of age, although the age range may extend from four months to seven years. The clinical course is typically acute with recurrences. A more protracted form of the disease may be seen following relapses and inadequate treatment. Signs include recurring fever, hyperesthesia, cervical rigidity, and anorexia. There may be a creeping gait, arching of the back with head held down, and crouched posture. Some dogs with protracted disease may show clinical signs of parenchymal involvement, such as ataxia, paresis, tetraparesis, or paraplegia. Hematological studies often reveal a peripheral neutrophilia with a left shift, increased erythrocyte sedimentation rate, and, in some cases, an elevated α-2 globulin fraction. CSF studies indicate increased protein and neutrophilic pleocytosis.

The cause of SRMA remains unknown. To date, no bacterial or viral infectious agents have been identified, although activated T cells have been found in some dogs, indicating these cells have had contact with some unidentified antigen.

The prognosis is guarded to favorable, especially in dogs with acute disease that are treated promptly using immunosuppressive doses of corticosteroids. Untreated dogs tend to have a remitting and relapsing course. Tipold recommends the following long-term therapy (e.g., for at least six months), especially in any dog that has had a relapse: prednisolone at 4 mg/kg/day, PO or IV initially. After two days, the dose is reduced to 2 mg/kg daily for one to two weeks, followed by 1 mg/kg daily. Dogs are re-examined, including CSF analysis and hematology, every four to six weeks. When signs and CSF are normal, the dose can be reduced to half of the previous dosage until a dosage of 0.5 mg/kg every 48 to 72 hours is attained. Treatment is stopped six months after clinical examination, CSF, and blood profiles are normal. In refractory cases, other immunosuppressive drugs, such as azathioprine (at 1.5 mg/kg PO every 48 hours), may be used in combination with steroids (e.g., alternating each drug every other day). Antibiotics are ineffective. Results of a long-term treatment protocol (up to twenty months) involving ten dogs with SRMA have been recently published. Eight of the ten dogs were without clinical signs up to twenty-nine months after the treatment was terminated. Long-term glucocorticosteroid treatment resulted only in mild clinical side effects, such as polyuria/polydipsia, polyphagia, and weight gain, which were reversible after the therapy was discontinued. It was noted that elevated serum and CSF IgA levels did not decrease to normal values during prednisolone treatment and were still slightly increased after the therapy was discontinued. Monitoring of CSF cell count in dogs with this condition was a sensitive indicator of success of treatment. In addition, older dogs with high
IgA levels in the CSF and frequent relapses seemed to require a longer duration of therapy and had a less favorable prognosis long term. Note that akitas, Bernese mountain dogs, and other breeds with immune-mediated polyarthritis may show similar clinical signs as animals with SRMA and have concurrent meningitis.

**Atlantoaxial Subluxation**

*Presentation and Pathogenesis*

The atlas (first cervical vertebra) and axis (second cervical vertebra) are bound together by ligaments that run from the dens of the axis to the atlas and the skull over the dens binding it to the floor of the atlas (the transverse ligament) and between the dorsal lamina of the atlas and the dorsal spinous process of the axis. The dens is a bony projection from the cranial aspect of the body of the axis and develops from a separate growth plate. Subluxation of the atlantoaxial junction is a relatively common problem and usually results from a failure of ligamentous support. Toy and small-breed dogs such as the chihuahua and Yorkshire terrier are at highest risk of the problem as a result of trauma. Onset of signs in dogs with the congenital form of the disease usually occurs in young dogs (less than two years of age), although problems can develop at any age. Signs can develop acutely or gradually, and waxing and waning of signs is often reported, presumably a reflection of instability at the atlantoaxial junction causing repeated injury to the spinal cord. Signs include neck pain (variably present), ataxia, tetraparesis, and postural reaction and conscious proprioceptive deficits with normal to increased muscle tone and myotatic reflexes in all four legs. In severe cases, animals can present with tetraplegia and difficulty breathing, and they may die acutely as a result of respiratory failure.

*Diagnosis*

Atlantoaxial subluxation can be diagnosed from survey radiographs of the cervical spine, although extreme care must be taken when restraining and moving dogs in which this disease is suspected. If the animal is sedated or anesthetized, the head and neck should be supported in slight extension to avoid further spinal cord injury. On lateral radiographs an increased space can be seen between the dorsal lamina of the atlas and the dorsal spinous process of the axis. In severe cases, malalignment of the bodies of the atlas and axis is clearly visible. The presence and size of the dens can be evaluated most accurately on VD views. If there is no evidence of subluxation on the lateral views, the neck can be carefully flexed to see if there is instability (the space between the dorsal lamina of the atlas and the dorsal spinous process of the axis should be evaluated). It is preferable to do this with fluoroscopy so that the movement can be monitored to prevent accidental iatrogenic subluxation. Recently, MRI evaluation of the spine and cord has proved beneficial in both the diagnosis and prognosis.

*Treatment*

**Conservative:** Dogs with mild signs can be treated conservatively by placing an external splint for at least six weeks. The splint must immobilize the atlantoaxial junction and so must come over the head cranial to the ears and go back to the level of the chest. The aim is to stabilize the junction while the ligamentous structures heal. The splint should be checked daily for signs of pressure sores by the owner and checked weekly by the veterinarian, with regular bandage changes if necessary. While often effective in the short term, the long-term efficacy of this approach is not known, and dogs treated in this way will always be at risk of repeated injury.

**Surgery:** Surgery is recommended in dogs with neurological deficits, although it can be associated with high perioperative morbidity and mortality. Dorsal and ventral approaches to the atlantoaxial junction have been described, but dorsal approaches are associated with a greater risk of causing spinal cord injury during surgery and a higher incidence of implant failure. Using ventral approaches, subluxation is reduced and the atlantoaxial articular surfaces are curetted to promote bone fusion. The two bones are fused using transarticular screws or Kirschner wires and a cancellous bone graft placed over the junction. In case of a traumatic injury or poor bone purchase, screws or Kirschner wires are placed in the body of the atlas and axis and the junction stabilized with polymethylmethacrylate cement. A neck splint is placed postoperatively while fusion occurs. This is a problematic area to repair surgically; bone quality is often poor, the bones are small, movement of the vertebrae may cause additional injury to the spinal cord, and the pharynx and larynx can be damaged during retraction. There is a risk of respiratory arrest and death in the perioperative period as a result of additional spinal cord injury, or inflammation of the upper airways secondary to retraction.
Prognosis
This is a serious disease, but dogs with mild deficits treated surgically have an excellent prognosis if they survive the 48-hour perioperative period. Although reported surgical success rates range from 50% to 90%, the majority report a mortality rate in the region of 20%, with the majority of deaths occurring either during or immediately after surgery. As with all spinal cord diseases, prognosis is worse in animals with severe and chronic neurological deficits. It has also been shown that prognosis is better in young dogs (< 24 months).

Cervical Disc Disease

Presentation and Pathogenesis
Cervical disc disease is a common problem in chondrodystrophoid breeds of dog such as dachshunds, Shih Tzus, and Pekingese. It also occurs frequently in beagles and cocker spaniels and can occur sporadically in almost any breed. Although thoracolumbar disc herniations have been reported in cats, cervical disc herniations are extremely rare. The intervertebral disc is composed of an outer fibrous portion (the anulus fibrosus) and a gelatinous center (the nucleus pulposus). With normal aging, the nucleus is slowly replaced by fibrocartilage, but in chondrodystrophoid breeds the nucleus ages prematurely and the nucleus matrix degenerates and mineralizes. As a result of these degenerative changes, affected dogs are prone to extrusion of the mineralized nucleus pulposus into the spinal canal, (Hansen type 1 disc herniations) causing spinal cord concussion and compression. The C2/3 disc is most commonly affected, with incidence decreasing further caudally in the cervical spine.

Onset of signs can occur from eighteen months of age, with a peak incidence between three and seven years of age. It is very unusual for a disc herniation to occur in dogs less than two years of age, as the predisposing degenerative changes have not occurred. The most common presenting sign is severe neck pain, as there is enough space within the cervical vertebral canal for herniation of disc material without compression of the spinal cord. The dog may adopt a stance with the head held down, neck rigid, and back arched as the weight is shifted to the pelvic limbs. Entrapment of nerve roots can cause a nerve root signature (holding up a thoracic limb and lameness). The neck pain is so severe that dogs avoid moving their head, and spasm and rigidity of the cervical musculature are easily palpable. Neurological deficits are less common but can occur when the spinal cord is sufficiently compressed; they include tetra or hemiparesis or -plegia, ataxia, and conscious proprioceptive and postural reaction deficits.

Diagnosis
Survey radiographs should be taken to identify degenerative changes typical of a disc herniation and to rule out other causes of the signs. Changes indicative of a disc herniation include narrowing of the intervertebral disc space, narrowing of the intervertebral foramen, and the presence of mineralized material within the vertebral canal and disc space. However, a definitive diagnosis cannot be reached with survey radiographs alone with adequate accuracy for surgery to be undertaken, and MRI, computed tomography, or myelography are used to identify the site of spinal cord compression. CSF analysis is performed concurrently to rule out an inflammatory disorder.

Treatment
Conservative: Dogs can be managed conservatively with strict cage rest for four weeks combined with pain relief using anti-inflammatory drugs, opiates, and/or muscle relaxants. Judicious use of anti-inflammatory doses of corticosteroids combined with appropriate cage confinement can be attempted if the pain is not responsive to nonsteroidal anti-inflammatory drugs (NSAIDs). Muscle spasm can also be responsive to gentle massage and hot packing of the neck. Administration of an H2 blocker such as famotidine may help to prevent the development of gastric ulceration. The aim of cage rest is to allow defects in the anulus fibrosus to heal, and resolution of pain does not mean that confinement should be discontinued. If this approach is successful, gradual reintroduction to controlled exercise can be attempted and the owners should be cautioned to prevent their pet from activities that involve jumping in the long term. Dogs should be monitored weekly and if the pain is unresponsive to conservative therapy, recurs, or neurological deficits develop, surgery should be recommended.

Surgery: Indications for surgery include unremitting or severe pain, recurrent pain, or neurological deficits. Once the site of disc herniation has been confirmed, a ventral slot is performed to remove the herniated disc material. Adjacent discs are fenestrated to prevent recurrence of the problem. Post operatively dogs are provided with pain relief and confined for four weeks (two weeks of strict confinement and then, if doing well, two weeks of increasing controlled exercise). Dogs are then gradually reintroduced to normal activity. If the dog has neurological deficits, postoperative care includes performing passive range of motion exercises, massage, hydrotherapy, and controlled exercise.
Prognosis
Prognosis for dogs treated conservatively is unknown. Prognosis for dogs treated surgically is excellent unless neurological deficits are severe.