

Caudal Occipital (Chiari-like) Malformation and Syringohydromyelia

A. Courtenay Freeman, DVM

Marc Kent, DVM, DACVIM (Small Animal and Neurology)

Simon R. Platt, BVM&S, MRCVS, DACVIM (Neurology), DECVN

BASIC INFORMATION

Description

The skull is made up of several flat bones that fuse together to encase and protect the brain. The bone of the back of the skull is called the *occipital bone*. Malformation or abnormal development of this bone compresses the back part of the brain, which is called the *cerebellum*. This compression causes the cerebellum to herniate (shift backward) and obstruct the hole in the skull where the brain joins the spinal cord. This hole is called the *foramen magnum*.

Obstruction of the foramen magnum alters the flow of cerebrospinal fluid (CSF). Normally, CSF is produced within the brain and flows around the brain and down the outside of the spinal cord. In caudal occipital or Chiari-like malformation, obstruction of CSF flow leads to the accumulation of fluid within the spinal cord. This fluid accumulation is referred to as *syringohydromyelia* (SHM).

The Cavalier King Charles spaniel is the most commonly affected breed of dog; however, other dogs, especially small-breed dogs, can also be affected.

Causes

Caudal occipital malformation is a congenital problem caused by abnormal development of the occipital bone of the skull. In the Cavalier King Charles spaniel, the condition may be inherited.

Clinical Signs

Onset of clinical signs may be sudden or may take several months to years to develop. Animals affected by this disease range from young to old.

Clinical signs often result from SHM and are related to spinal cord dysfunction. Signs include neck pain, weakness, incoordination, and muscle loss over the shoulders. Scoliosis (abnormal curvature of the spine) and twisting of the head can occur. Affected animals may excessively scratch at their head and neck, especially when excited. Facial muscle paralysis and imbalance are occasionally observed. Seizures have also occurred in some affected animals, but they may be unrelated to this malformation.

Diagnostic Tests

A neurologic examination, routine laboratory tests, and x-rays are often recommended to rule out other diseases that produce

similar signs. Confirming the diagnosis usually requires magnetic resonance imaging (MRI). MRI can reveal distortion of the brain structures, obstruction of CSF flow at the foramen magnum, and the presence of SHM.

TREATMENT AND FOLLOW-UP

Treatment Options

Medical and surgical treatments exist. In most cases, medical therapy is pursued initially and is aimed at decreasing CSF production and providing pain relief. Corticosteroids can be used to reduce CSF production. Occasionally, diuretic drugs may also be needed. Pain medications, such as gabapentin or tramadol, may be used in animals that experience discomfort or painful episodes.

Surgical therapy involves enlarging the foramen magnum in an attempt to restore CSF flow. When the obstruction is relieved, the CSF does not continue to accumulate within the spinal cord. This surgery may not eliminate fluid that has already accumulated, so some clinical signs may not improve. Surgery is most often considered for dogs with severe signs or signs that worsen despite medical therapy.

Follow-up Care

Follow-up visits are scheduled frequently at first, to monitor the success of medical or surgical treatment. Recheck appointments may eventually be decreased to every 6 months if the animal responds to treatment and has no complications. A recheck MRI may be helpful to evaluate the success of treatment and to identify any progression of the disease.

Prognosis

Prognosis depends on the severity of clinical signs and the extent of structural damage to the brain and spinal cord. Many mildly affected dogs can be successfully treated with medical therapy. In some dogs clinical signs improve after surgery, but surgery is not always successful, and recurrence of signs is possible. In some dogs, surgery may halt worsening of the signs and allow medical therapy to be more effective.