

Chiari-like Malformation and Syringomyelia

Chiari-like malformation (CM) is a genetic neurological condition in dogs characterized by abnormal growth of bone in the back of the skull, basically making the skull too small for the brain.

Pressure resulting from this tight space affects the flow of cerebrospinal fluid. This can result in fluid-filled pockets forming within the spinal cord, a secondary condition known as syringomyelia (SM). Unfortunately, as the fluid pockets expand over time, they ultimately damage the center of the spinal cord.

Signs

Typically, signs become apparent once Chiari-like malformation results in syringomyelia. The most obvious symptom is neuropathic pain, or the abnormal processing of sensory input by the nervous system. The altered sensation usually occurs at the back of the head and neck, which is why you may find your dog constantly scratching, often without making contact and to no avail, known as *phantom scratching*.

Symptoms vary among breeds and individuals, but these are the most common signs in dogs:

- Phantom scratching
- Scratching at ears, neck, or back of head
- Rubbing at face
- Sensitive when touched around back of neck or shoulders
- Pain when active, excited, passing stool, or simply wearing collar/harness
- Change in behavior (lowered head, less playful, or withdrawn)
- Limb weakness or incoordination
- Scoliosis (curved spine)

Causes

The cause of Chiari-like malformation and secondary syringomyelia is a genetic malformation, or birth defect, and thought to be hereditary. These conditions are generally rare, but toy breeds are overrepresented, especially Cavalier King Charles Spaniels and Brussels Griffons.

Virtually all Cavalier King Charles Spaniels have Chiari-like malformation to some degree. However, it is important to note that not all dogs with one or both conditions will show symptoms.

Diagnosis

Your neurology care team will consider breed, age, history, and symptoms during physical and neurological exams, but the only way to definitively diagnose CM/SM, rule out other health problems, and decide on a treatment course is with magnetic resonance imaging (MRI).

Treatment

The goal of treatment is to manage neuropathic pain and neurologic deficits through pharmaceutical or surgical approaches.

Medical treatment involves some combination of pain management, anti-inflammatories, and medications that reduce the production of cerebrospinal fluid. Unfortunately, studies show only about 10% of dogs improving from medical management alone long-term. This condition is often progressive, and surgery is recommended because the risks are low and the procedure corrects the original cause.

Surgical treatment involves removing malformed bone at the back of the skull to make more space for cerebrospinal fluid to flow out of the brain. Surgery can be much more effective for the right patient, with studies showing over 80% of patients improving immediately, with many cases doing well long term. However, it is important to note that surgery should be performed before symptoms become severe, as surgery tends to slow progression but does not always reverse damage that has already occurred.

Prognosis

Chiari-like malformation and the subsequent development of syringomyelia are congenital diseases that cannot be prevented or cured. However, through early diagnosis, progression can be slowed, pet parents can be better prepared, and passing those traits to future generations can be stopped.

At Southeast Veterinary Neurology (SEVN), we offer a heavily discounted screening package for at-risk breeds that do not currently have symptoms to identify the conditions as early as possible. This is particularly useful in responsible breeding.

Most importantly, advanced screening will help provide the best achievable outcome for your dog. Visit our website for details.



Learn more about
Chiari-like
Malformation

