Chiari-like malformation (CM, as referred to further on) is a condition where the back of the skull/ cranial cavity is too tight for the brain matter which results in the cerabellum and often also the brain stem pushing to or through the opening in the back of the skull (foramen magnum).

CHIARI–LIKE MALFORMATION AND SYRINGOMYELIA

Summary: Finnish Russian Toy Club/Breeding committee
Pictures: Nina Saksman/Aisti

Syringomyelia (SM, as referred to further on) is the development of fluid filled syrinx or syringes in the spinal cord due to the abnormal flow of the spinal fluid. The biggest factor in the development of syringomyelia is CM.

These diseases can greatly reduce the quality of the dog's life by causing neuropathic pain and motor dysfunction.

WHEN THE BRAIN DOESN'T FIT THE SKULL

CM is condition of multiple problems, such as abnormal development of the skull and craniocervical junktion, the cerabellum and often also the brain stem pushing to or through the opening in the back of the skull (foramen magnum) and over all tightness in the back of the skull.

Both CM and SM are somewhat typical in small and toy sized dogs, but it is most famously known as a disease of the Cavalier King Charles Spaniels. CM also occurs as a similar condition in humans (Chiari malformation) hence the name Chiari-like malformation.

SYRINGOMYELIA

In SM the flow of the spinal fluid is disrupted which results in spinal chord lesions/fluid filled cavities called syringes. Other factors, such as trauma, tumor, vertebrae disk or inflammation can also cause these syringes if the flow of the spinal fluid is abnormal. Most common cause however is CM.

Usually syringes appear in the cervical vertabrae area, but research of the Cavaliers show that syringes can appear throughout the whole spine. However, all dogs with SM have syringes at least in the cervical vertabrae.

Although heredity of SM has been studied, the method has not been clarified. It is most likely polygenic and appears to have moderately high heritability.

CLINICAL SYMPTOMS OF SM

There are many symptoms that can sometimes be hard to notice or connect to SM.

- scratching or sensitivity of either side of the shoulder
- sudden squels
- ataxia and clumsiness
- paralysis of the facial nerves
- scoliosis
- torticollis (scoliosis of the neck)
- epileptic seizures
- rubbing of the head and face
- separation anxiety
- hypersensitivity to touch
- aggressiveness
- fear of strangers

Location of the pain is usually in the neck area, but it may be difficult to find. Scratching may appear in the painful areas, typically on one side only. It doesn't always involve contact to the skin (scratching the air without touching the body). Excitement can be a trigger to this action.

Torticollis

Sensitivity of a painful area can result in the dog not allowing itself to be touched in the area. Wearing a collar or clothes can cause pain or unpleasant sensation. The pain can be long lasting even after a slight touch.

Some dogs might not show any symptoms of neuropathic pain. Size and shape of the syrinx affects on the severity of the symptoms.

SM can also be the cause of behavioral issues such as separation anxiety and aggression or fear toward strangers. Dogs with the most pain handle stress the worst and are unable to calm down properly.

However, not all symptoms can be explained with SM since some of them occur with dogs diagnosed only with CM. On the other hand, CM doesn't always cause visible symptoms so it may go unnoticed until it may lead to SM and the related symptoms.

DIAGNOSIS AND TREATMENT

Diagnosis for either or both can only be confirmed with magnetic resonance imaging (MRI). Finding a veterinary clinic or hospital with the appropriate equipment can be hard and the examination is usually expensive.

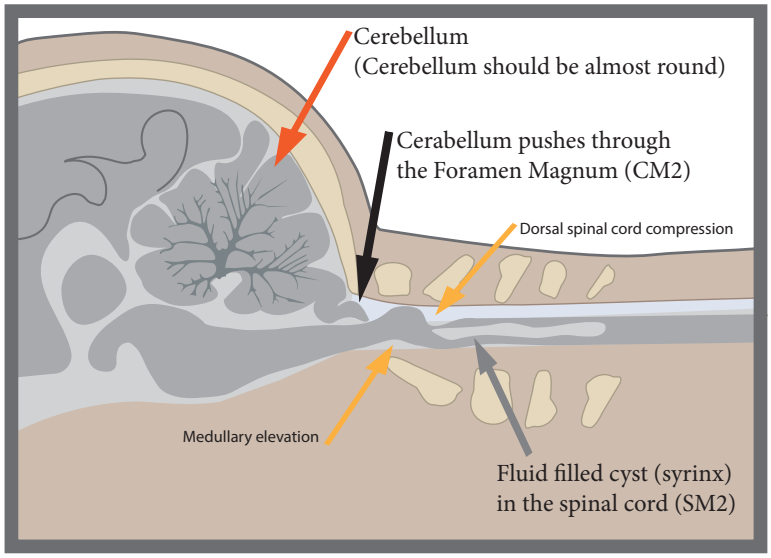
SM is a progressive condition. There is no definitive cure for CM or SM. No medication is scientifically proven to fully eliminate the pain. Usually the dog is medicated with at least painkillers for the rest of it's life. Surgery can be used to slow down the progress in some cases but there is no evindence of it's benefit.

OCCURENCE

CM and SM are hereditary in Cavaliers. Almost every cavalier is affected by CM and almost half (46%) of the symptom free dogs have SM as well. CM and SM are quite common in Griffon Bruxellois. There are multiple reported cases in Yorkshire Terriers, Toy Poodles, Chihuahuas, Pugs and French Bulldogs.

Several Russian Toys in Finland have been scanned in the past few years with devastating results. Dogs without any symptoms and with severe symptoms have been diagnosed with either or both CM and SM. We have an on-going study with University of Helsinki "Persistent fontanelles in Association with craniocervical junction abnormalities, syringomyelia, and ventricular volume" where they needed samples from toy sized dogs under 5 kg.

The Finnish Russian Toy Club will publish more information on the matter when official results are available and the study in the University of Helsinki progresses.



Chiari-type malformation and Syringomyelia classification:

	central canal dilatation/syringomyelia:
SM0	**normal, central canal dilatation under 1,0 mm (no central canal dilatationunder noticeable)**
SM1	**central canal dilatation 1,0-1,9 mm**
SM2	**central canal dilatation 2 mm or higher, also syrinx formations outside the central canal (regardless of the diameter of the central canal) or precursor of the syrinx (presyrinx)**
	Chiari-type malformation:
CM0	**normal cerebellum posterior cranial fossa**
CM1	**indentation on the back edge of the cerebellum, but cerebrospinal fluid visible between the back of the vermis and the foramen magnum**
CM2	**herniation of the cerebellum into or through the foramen magnum**

Sources:
https://helda.helsinki.fi/bitstream/handle/10138/154688/lisensiaatin_tutkielma_Johanna_Forsgard_2015.pdf?sequence=1&isAllowed=y
https://helda.helsinki.fi/bitstream/handle/10138/303821/Lisensiaatin%20tutkielma_Juulia%20Martikainen.pdf?sequence=2&isAllowed=y
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