

Etiopathogenetic mechanism in dogs with syringomyelia

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Abstract: Syringomyelia (MS) is a condition characterized by the development of cavities in the parenchyma of the spinal cord (sirinx). Cavalier King Charles Spaniel (CKCS) dogs have a high incidence of Chiari malformation. The breed was born in 1928, from dogs of the King Charles Spaniel breed, a favorite dog of royal and noble families, with the aim of recreating a dog similar to the one present in the portraits of King Charles II, during the restoration period. The CKCS breed is native to the United Kingdom, being composed of small brachycephalic specimens - toy, a distinctive feature of the breed being the flattened and miniaturized appearance of the head (Rusbridge & Knowler, 2003).

Syringomyelia is a topical and real interest topic in canine neuropathology, representing the topic of numerous researches, both due to the many unknowns of the evolution of the diseases, and the fact that at present there is no effective treatment.

Keywords: Central nervous system, genetic predisposition, CT exam, behavioral changes, nervous symptoms

1. Introduction

In dogs, the etiology of syringomyelia is not fully known, but in the case of Cavalier King Charles Spaniel and Griffon de Bruxelles, syringomyelia may occur as a result of gene modification. BMP 3 (bone morphogenetic protein 3) is the gene responsible for regulating the harmonious development of the skull and spinal cord, so that the extreme brachycephalic conformation of these breeds is associated with Chiari malformation and syringomyelia. Two chromosomal loci were identified - CFA 22 and CFA 26, which are associated with the presence of a reduced volume of the caudal cranial fossa and with the modified orientation of the caudal cranial fossa, characteristic of Chiari malformation and syringomyelia [1] (Miller & Zachary, 2017), [2] (Schoenebeck, et al., 2012).

2. Receptivity

Syringomyelia (MS) are commonly diagnosed in Cavalier King Charles Spaniel and Griffon de Bruxelles breeds, but are also found in other brachycephalic breeds, including: Chihuahua, Maltese Bichon, Pomeranian, Pug, French Bulldog, Yorkshire terrier, Boston terrier, but and their mestizos (Dewey, et al., 2005) (Rusbridge, 1997).

Syringomyelia are rarely diagnosed in cats, however recent studies have indicated the presence of MS in cats, especially in brachycephalic breeds.

In the literature, syringomyelia has rarely been described in other animal species, especially in horses and calves, in most cases being a congenital disease, not being correlated with MC (Chiari-like malformation).

3. Etiopathogenesis

One of the most common causes of syringomyelia (MS) is Chiari-like malformation (MC), secondary to volume mismatch between the caudal cranial fossa

Received: 7 December 2021

Accepted: 22 February 2022

Published: 28 June 2022

DOI:10.52331/cvj.v27i1.33



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(too small) and brain tissue (too large), altered CSF (cerebrospinal fluid) circulation, and subarachnoid space compression (Freeman, et al., 2014) (Rusbridge, 2014). In addition to MC, syringomyelia can also be associated with many pathologies that cause CSF obstruction. However, it is unclear why only some dogs diagnosed with Chiari-like malformation (MC) develop MS and others do not. Other causes besides MC that can lead to syringomyelia are CSF obstruction (neoplasms), trauma, constrictive lesions of the spinal cord or space-occupying processes in the spinal cord. In a study conducted in 2013 [3] (by Driver et al.) in dogs with and without MS, pulsation cerebellar was assessed during the cardiac cycle using magnetic resonance imaging. These findings support theories of the pathogenesis of syringomyelia secondary to Chiari malformation in human medicine, highlighting the similarities between canine and human patients.

There can be many factors that influence the pathogenesis, among which we list intracranial hypertension, blood-brain barrier disturbances, imbalance between CSF production and absorption, or insufficiency of extracellular fluid absorption or drainage (Hemley, et al., 2012).

5. Physiopathology of syringomyelia

Due to the underdeveloped caudal cranial fossa, its small volume cannot fully encompass brain tissue. This abnormality leads to the herniation of the cerebellum, thus obstructing the flow of cerebrospinal fluid (CSF) through the foramen magnum (FM), and the CSF pressure exerted on the CNS (central nervous system) is inconsistent CSF flow disturbance as well as pressure variations seem to play an important role in development of syringomyelia. Cerebellar herniation appears to be a component of MS. (Fig.2). It is common in cases of MS, but the presence or size does not anticipate the formation of MS. An explanation by which herniation cerebellar contributes to the pathogenesis of MS is that obstruction of CSF channels occurs in the foramen magnum, but there must be other predisposing factors. (Cerdeira-Gonzalez, et al., 2009b) (Lu, et al., 2003).

Another important factor included in the pathogenesis of MS is CSF circulation. Changes in speed, turbulence and disturbances in CSF circulation can lead to the formation of intramedullary cavities. A high rate of CSF flow recorded in the foramen magnum along with a lower rate of CSF flow in the cervical vertebrae C2-C3 are considered predisposing factors in the formation of MS.

Intracranial pressure is higher than in the cervical portion of the spinal cord, so when there are rapid increases in pressure, CSF converges to the cervical region. Inside the spinal cord, the pressure tends to increase faster in the lumbar region than in the cervical area, further favoring CSF movement toward the cervical portion of the spinal cord. (Rusbridge, et al., 2000) (Kirberger, et al., 1997)

Due to the direct link between MS and CSF circulation disturbances, the size of the cerebral ventricles correlates with the size of the medullary cavities. CKCS dogs diagnosed with MC and MS have smaller jugular foramen (jugular hole) compared to CKCS patients diagnosed only with MC. Because of this, the venous shaft at the level of the jugular hole is reduced and associated with a small cranial base, leading to increased venous pressure and reduced absorption of CSF, predisposing factors in the formation of intramedullary cavities. (Rusbridge, et al., 2009a) (Schmidt, et al., 2012).

Another cause of reduced CSF absorption is low sinus volume in CKCS patients diagnosed with MC and MS. Reduced venous sinus volume leads to increased intracranial pressure and secondary to improper CSF reabsorption.

3. Discussions

The pathophysiology of syringomyelia is still unclear, several theories are exposed in the literature. However, the full mechanism is not fully understood, and has been the subject of intense research in recent years. It is important to note that the tubular formations in the spinal cord parenchyma, which does not involve the central medullary canal, are defined as syringomyelia (syrinx), while hydromyelia is defined as a dilation of the central canal of the spinal cord. These two cavities can communicate with each other, but it is difficult to highlight this communication. In the early stages, the cavities are located dorsally and laterally by the central medullary canal, in the gray matter. The medullary tissue around the cavities is edematous. If syringomyelia is causing signs and symptoms that interfere with the life of animal, it is recommended a treatment based on Gabapentin at 10 mg/kg orally every 8 to 12 h as a primary treatment for several symptoms like: neck and back pain on palpation, abnormal scratching, episodes of sudden vocalizing or in advanced stages is necessary surgery. The goal of surgery is to remove the pressure the syrinx places on your spinal cord and to restore the normal flow of cerebrospinal fluid. This can help improve the symptoms and nervous system function (Fig. 1).

4. Conclusion

Overall, the prognosis for CM/SM-affected dogs depends on the severity of clinical signs and on the response to medication. Chiari-like malformation and syringomyelia is a progressive condition in those dogs that are affected clinically. Some dogs will need constant dose adjustments to adequately treat their symptoms. Unfortunately, some dogs afflicted with severe and disabling pain do not respond to medical management and are surgical candidates.



Fig.1 Differences between a healthy dog and a dog with Syringomyelia (<https://www.frontiersin.org/articles/10.3389/fvets.2018.00280/full>)

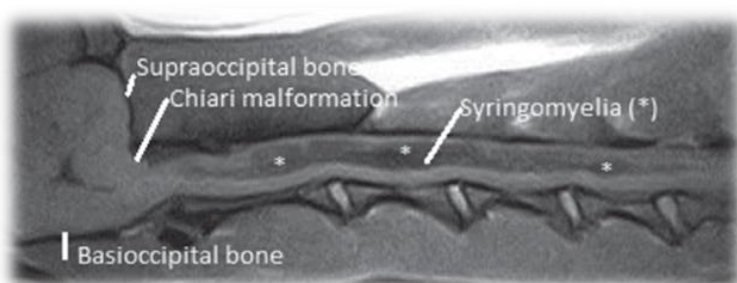


Fig. 2 Chiari malformation and Syringomyelia seen on a CT image (<https://www.vin.com/apputil/content/defaultadv1.aspx?id=5328340&pid=11349&print=1>)

References

1. Couturier, J., Rault, D., & Cauzinille, L., 2008. Chiari-like malformation and syringomyelia in normal cavalier King Charles spaniels: a multiple diagnostic imaging approach. *Journal of Small Animal Practice*, 49, 438–443.
2. Teză de Doctorat, Cucoş C.A., Diagnosticul Malformației de tip Chiari și a Siringomieliei la Câine, București, 2019.
3. Dewey, C., Berg, J., Barone, G., Marino, D., & Stefanacci, J., 2005. Foramen magnum decompression for treatment of caudal occipital malformation syndrome in dogs. *Journal of the American Veterinary Medical Association*, 227(8), 1270-1275.
4. Driver, C., De Risio, L., Hamilton, S., Rusbridge, C., Dennis, R., & McGonnell, I., 2012. Changes over time in craniocerebral morphology and syringomyelia in cavalier King Charles spaniels with Chiari-like malformation. *BMC veterinary research*, 8(1), 215.
5. Freedman, D., 2011. Preliminary Morphometric Evaluation of Syringomyelia in American Brussels Griffon Dogs. *Journal of Veterinary Internal Medicine*, 25(3).
6. Rusbridge, C., & Knowler, S., 2006a. Coexistence of occipital dysplasia and occipital hypoplasia/syringomyelia in the cavalier King Charles spaniel. *The Journal of small animal practice*, 47(10), 603-606.
7. Dewey C, Berg J, Barone G, et al. Foramen magnum decompression for treatment of caudal occipital malformation syndrome in dogs. *J Am Vet Med Assoc*. 2005;227:1270–1275.