Artykuł przeglądowy *Review*

Clinical anatomy of syringomyelia and Chiari malformation in dogs

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Summary

Syringomyelia (SM) is a rare disorder characterized by the development of fluid filled cavities (syrinxes) in the spinal cord parenchyma which occurs secondarily to an obstruction of cerebrospinal fluid (CSF) outflow at the foramen magnum level. The precise pathophysiology of SM is still not fully understood. The most common predisposing cause in dogs is a Chiari-like malformation (CM). This disease is defined as a developmental failure of the occipital bone leading to overcrowding of the caudal cranial fossa. Abnormally small bony structures contain rhomencephalon structures (cerebellum and medulla oblongata) which are unchanged in size. This "mismatch" causes caudal displacement of encephalon structures via the foramen magnum, altering the cerebrospinal fluid flow. CM is thought to be associated with a higher grade of brachycephaly. The SM/CM complex is very common in Cavalier King Charles Spaniel (CKCS) breed.

Keywords: Chiari malformation, syringomyelia, occipital hypoplasia, brachycephaly

Chiari-like malformation syndrome, or simply Chiari malformation (CM), is a morphological abnormality of the caudal fossa of the cranial cavity. It is described as a mismatch in volume between encephalon structures and caudal fossa, which leads to displacement of rhombencephalon parenchyma towards the vertebral canal (6). These changes are sometimes described as the cerebellar herniation (7, 10, 24, 41). Consequently, some disturbances occur at the level of the foramen magnum in the flow of cerebrospinal fluid (CSF). Aleteration of CSF flow dynamics may lead to accumulation of tissue fluid in the spinal cord and eventually cause the development of cavities (syrinxes) in its parenchyma (21, 32, 33, 40, 47). This state is known as syringomyelia (SM) (8, 11, 22, 34, 36, 38). The above mentioned disease entities are mainly diagnosed in people. They are more and more frequently diagnosed due to the development of the techniques of diagnostic imagining. One of them is MRI, which allows the detection of relatively small changes with a symptomless course (14, 19). Despite numerous clinical observations and experiments performed on animals, the pathogenesis of SM is not fully explained (26, 27, 49). These disturbances are also observed in dogs, mainly in brachycephalic breeds and the so called toy breeds. They were described, among others, in Cavalier King Charles Spaniels (CKCS), Brussels Griffons and Yorkshire Terriers (4, 23, 40).

The aim of the work is a review of the research results concerning the influence of morphological traits on the pathogenesis of the above mentioned diseases and the presentation of the current knowledge about the hydrodynamics of the cerebrospinal fluid.

An outline of the morphology of the cranial cavity in dogs from the perspective of syringomyelia and Chiari malformation

The shape of the cranial cavity and the internal surface of bones forming it are "sculptured" by the respective structures of the brain. Three main structures can be distinguished in its area, gradually declining towards the back: rostral cranial fossa (*fossa cranii rostralis*), middle cranial fossa (*fossa cranii media*) and caudal cranial fossa (*fossa cranii caudalis*). Their "inhabitants" are respectively: olfactory bulbs (*bulbus olfactorius*) and the most frontal descriptive elements of the hemispheres, in the next – the remaining part of the telencephalon together with diencephalon and mesencephalon, and in the last part – metencephalon together with myelencephalon, i.e. rhombencephalon s. medulla oblongata (Fig. 1). The caudal cranial fossa is located over the basilar part of the occipital bone

(*pars basilaris ossis occipitalis s. basiooccipitale*). On the sides it is limited by the petrous parts of the temporal bones (*partes petrosae ossis temporalis*), and from the back by the squamous part of the occipital bone (*squama occipitalis*). The bottom vault is formed by the osseous cerebellar tentorium (*tentorium cerebelli osseum*), and its symbolic frontal limit is the line running from the mentioned structure to the rostrally situated dorsum sellae. On the ventral surface of the petrous part (*facies ventralis partis petrosae*) there is a petrooccipital fissure (*fissura petrooccipitale*). That structure is in direct contact with the jugular foramen (*foramen jugulare*) located on the external surface of the base of the skull (Fig. 2). Through that foramen passes the internal carotid artery (*a. carotis interna*) and internal jugular vein (*v. jugularis interna*) which is the main drainage of blood from the brain. The back border of the caudal cranial fossa is formed by the foramen magnum. From the top, that structure is limited by the squamous part of the occpital bone and on the sides by lateral parts of the occipital bone (*partes laterales ossis occipitalis s. exoccipitale*). The lower border of the foramen magnum is formed by the basilar part of the above mentioned bone. The foramen magnum marks the arbitrary border between the medulla oblongata and spinal cord. The developmental disturbances concerning the presented osseous structures forming the caudal cranial fossa and foramen magnum results in the misfitting of the brain's anatomical elements and appearance of the discussed disease entities.

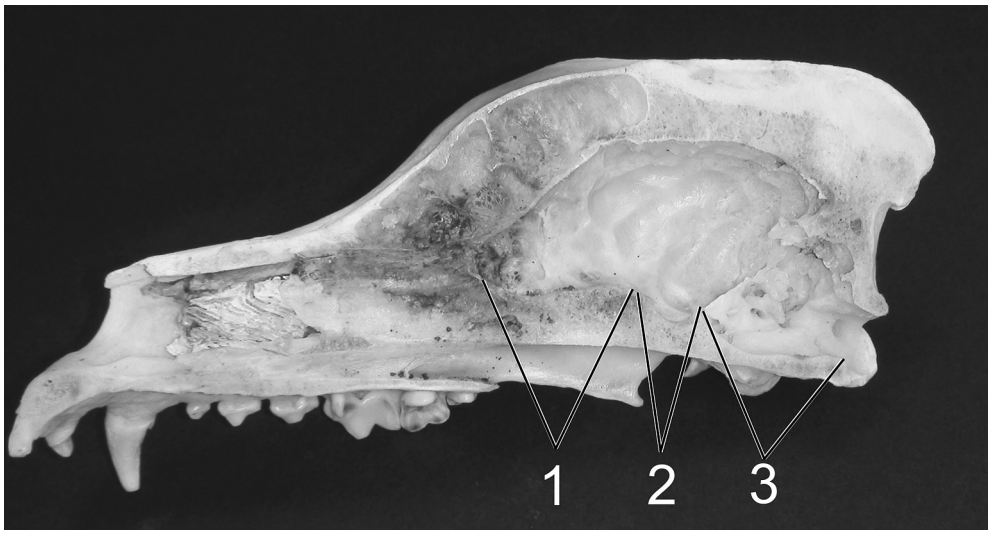

Fig. 1. Medial aspect of sagittal section of a dog skull. 1 – rostral cranial fossa, 2 – middle cranial fossa, 3 – caudal cranial fossa

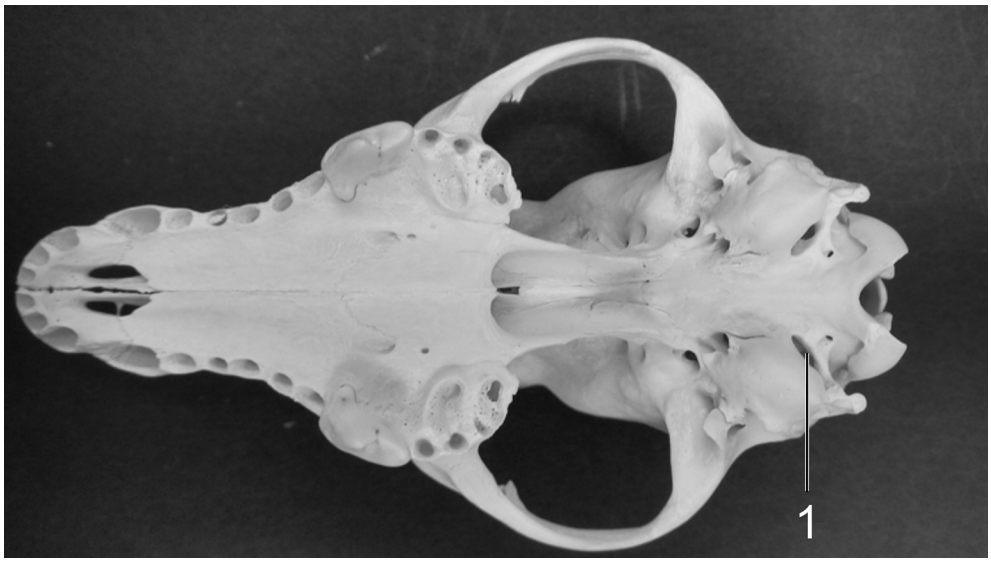

Fig. 2. Ventral aspect of a dog skull. 1 – jugular foramen

Chiari and Chiari-like malformation

In 1981 an Austrian pathomorphologist Hans von Chiari published a monographic description of human brain developmental disturbances which were characterized by the displacement of the elements of rhombencephalon towards the vertebral canal. He classified the observed changes as type I, II and III disturbances, and five years later he also described type IV (13). The division was based on the analysis of the degree of herniation of particular rhombencephalon structures into the vertebral canal and the differences connected with it in the course of the disease. It should be stressed that the gradation of pathology within types I–III worked out by Hans von Chiari was universally accepted and is still used. However, many authors do not accept type IV. In 1894, German pathomorphologist Julius Arnold described a case of an infant with the type II defined by Chiari. On account of that, for a long time the above discussed pathological state was described as the Arnold-Chiari syndrome (37, 50).

The more poorly expressed type I characterizes the displacement of medulla oblongata into the vertebral canal and the herniation of the cerebellar tonsils (*tonsilla cerebelli*) into the foramen magnum. It appears mainly in 20-30-year-old people, thus it is called the Chiari malformation of adults (2, 25). Symptoms indicating type II malformation are present already at birth. Anatomically a herniation of the vermis of the cerebellum, pons and fourth ventricle (*ventriculus quartus*) into the foramen magnum and the medulla oblongata into the vertebral canal are observed. Most often that

pathology coexists with other developmental disorders, such as rachischisis (13, 37). Type III is characterized by a displacement of the entire cerebellum into the foramen magnum and vertebral canal. The described by Chiari type IV consists in cerebellar hypoplasia with rhombencephalon located within the posterior cranial fossa which topographically corresponds to caudal cranial fossa in animals (13).

It is assumed that the most often observed type I, also called classic, results from the hypoplasia of the basilar part of the occipital bone which consequently causes the volume decrease of the posterior cranial fossa. It leads to the "overfilling" of the space of the fossa and squeezing of the cerebellum into the foramen magnum (1, 25, 29, 30). Changes similar to those observed in the type I malformation in humans are also noted in dogs, thus in veterinary medicine they are defined as the Chiari-like malformation. However, the name is doubtful because the lesions noted in dogs do not fully match those observed in humans in particular types of the disease. In dogs anatomical changes within the occipital bone predominates thus that disease has also been described as the caudal occipital malformation syndrome or occipital hypoplasia (4, 38, 39).

The breed particularly predisposed to the discussed lesions is the Cavalier King Charles Spaniel. Some reports can be found in literature describing that disease in other breeds of dogs. Rusbridge et al. diagnosed it in 34 out of 56 examined Brussels Griffons (40). According to that author the breeds which are equally endangered with the appearance of that syndrome are: Maltese, Chihuahua, Miniature Poodle, Shih Tzu, Pomeranian, Boston Terriers and Pekingese dogs. Cagle described that syndrome in a 7.5-year-old Yorkshire Terrier (4). In their report, Park et al. described the CM cases in a Poodle and Maltese (32). Most of the mentioned breeds belong to the brachycephalic morphotype. They are characterized by a specifically shaped head skeleton with a clearly shortened viscerocranium, hypoplasia of the jaw, strongly curved zygomatic arch and a dome-like bulge of bones limiting the neurocranial vault. In dogs of that morphotype the so called brachycephalic syndrome is very often diagnosed. The osseous changes are often accompanied by other disorders, such as rotation of the teeth, narrowing of the anterior nostrils, bending the long axis of the nasal meatus and nasal conchas, long soft palate, abnormalities in the structure of the larynx and trachea and respiratory problems (43). The so called occipital dysplasia is often observed. In the course of it there appear some disturbances in the ossification of the squamous part of the occipital bone, which lead to the dorsal increase of the foramen magnum (33, 48). In their research on the morphology of the foramen magnum in dogs, Watson et al. observed that the increase of the skull index (the more brachycephalic skull) is accompanied by the increase of the frequency of the appearance of occipital dysplasia (48). They also noted that the characteristic dorsal incisure increasing the foramen magnum in all cases was covered by a connective tissue layer. At present, it is still debated whether dorsal notch is a pathology or only an expression of an anatomical variability within the foramen magnum (16).

Apart from the above described changes, the base of the skull becomes shortened in the brachycephalic dogs (40). This is probably connected with the premature closure of the junctions between basilar parts of the presphenoid, basisphenoid and occipital bones (44). At the same time some changes take place within the cranial vault. A compensatory growth of the parietal bone which forms space for the developing brain can be observed. As a result it becomes rounded and clearly widened (40). Similar changes in the spatial arrangement of the cranial bones were described in humans with the Chiari syndrome (35).

Rusbridge and Knowler described a simultaneous hypoplasia and dysplasia of the occipital bone in two males of the CKCS breed (38). In both those cases a development of SM was observed, but surprisingly the syndrome occurred at a relatively old age, 8 and 10 years, respectively. This is remarkable because it is assumed that the SM which develops secondary to CM usually appears before the end of the seventh year of life (39). While describing the cases of two CKCS males the authors present the opinion that the connective tissue covering the dorsal incisure of the foramen magnum fascilitates the flow of the cerebrospinal fluid, delaying the appearance of the disease symptoms. An interesting hypothesis proposed by Shaw et al. should be mentioned here (45). The authors state that similarly as in humans the modeling of cranial bones in dogs also takes place after the closure of sutures. They suppose that it is one of the causes of shaping the dorsal incisure. Additionally, a clear thinning of the squamous part of occipital bone is frequently observed. It was assumed that there is bone resorption, which is an expression of the cranial compensatory changes for creating space for the developing brain.

There are reports which show that the cerebellar hernia was diagnosed in 95% of the examined CKCS population (7). Thus, some attempts were undertaken to explain the exceptional predisposition of that breed to CM. It was found that in comparison with the mesaticephalic dogs the caudal cranial fossa in CKCSs is clearly more shallow. Also there often appear some anatomical disorders within the squamous and basilar part of the occipital bone (5). In other representatives of the brachycephalic morphotype such a relationship was not observed (42). It has been reported in other publications that differently from other brachycephalic breeds dogs of the CKCS breed have a very wide neurocranium in relation to its length (42, 43). Moreover, the dimensions of their brains are comparable to respective structures in Labradors which causes the "misfitting" of the soft structures located in the caudal cranial fossa (9). That

phenomenon is characteristic only for the CKCS breed. Other brachycephalic dogs have the brain proportional to the cranial cavity volume. The author also noted that the caudal cranial fossa in the CKCS dogs contains the higher per cent part of the brain as compared with Labradors or small breed dogs. He suggested that the CM pathomorphology in dogs can also be connected with the morphology of the rostral cranial fossa. It is assumed that in the process of domestic dog miniaturization there was a simultaneous decrease of the size of both the skull and the brain. That mechanism in the CKCS dogs was probably disturbed in such a way that only the skull became smaller.

The results of the investigations by many authors point to the fact that the CM is a multifactorial disease with its pathogenesis not as yet fully researched.

Syringomyelia

The first descriptions of the disease date from the 16th century. The term syringomyelia was used for the first time by Olivier d'Angers in 1824. At the beginning it was assumed that the described syrinxes were the widening of the spinal cord central canal. Some trials were undertaken at systematizing the observed changes. They tried to base them, among other things, on the type of fluid filling the spaces or on the type of cells comprising their lining. In the past the discussed lesion was described as hydromyelia or syringohydromyelia. At present, the name syringomyelia covers all clinical states in which syrinxes develop in the spinal cord and contain tissue fluid. This classification does not include syrinxes associated with tumors (37).

Pathogenesis of SM is a complicated process not fully explained yet. It is commonly known that its development is conditioned by many states, such as developmental disorders, traumas, cerebral meningitis and neoplasms within the spinal cord (3, 18, 28, 31, 36, 47). There are several theories which aim at explaining the mechanisms leading to the appearance of the disease. In 1950, a classic, so called, "water hammer" theory was proposed by Gardner. He based it on the axiom that the cerebrospinal fluid flow takes place in accordance with the cardiac rhythm. The cited author suggests that in the cases of hampering the drainage at the level of the foramen magnum, each myocardial contraction causes the pushing ("water hammer") of a portion of fluid into the central canal. Consequently, there is a gradual dilation of the central canal, fluid penetration of the nervous tissue of the spinal cord and formation of syrinxes. Further investigations revealed that the majority of patients with SM do not have a patent connection between the fourth ventricle and the central canal. According to Rusbridge et al. such communication is much more often observed in dogs (37). However, on the basis of the MRI investigations, the authors claim that there is no proof allowing a statement that all the syringomyelic cavities originate from the central canal. The remaining theories, e.g. the theories by Williams (1976) and Oldfield (2001) are also unable to explain the mechanism responsible for the development of SM. At present, Greitz's theory of an intramedullary pulsation pressure is accepted (11, 12). That theory enables the description of the pathophysiology of SM irrespective of its etiology. As it was already mentioned, the development of SM is always secondary to the disturbances in the cerebrospinal fluid flow. It is constantly produced in the choroid plexus of cerebral ventricles and capillary vessels. It is generated as a result of filtration and active transport. So far it has been assumed that the increased pressure of the cerebrospinal fluid inside the subarachnoid space causes its pushing into the existing syringomyelic cavity. This theory is inconsistent, because it is known that the increased pressure outside the existing space should crush it. Greitz's theory assumes that the mechanism leading to SM is the increase of pulsation pressure inside the spinal cord as compared with the lower pressure in the surrounding subarachnoid space. An assumption was made that syringomyelic cavities appear as a result of repeated mechanical dilatation of the spinal cord. In the situation in which the subarachnoid space becomes narrowed there is a decrease of the transmission of the systolic wave of the cerebrospinal fluid in the caudal direction. At the same time there is the transmission of the fluid pulsation pressure to the spinal cord tissue. When an increase of pressure inside the spinal cord and its decrease in the surrounding subarachnoid space transpires the cord becomes dilated directly below the narrowing (12, 17). As a result of the reflection of some of the pulsation pressure of the cerebrospinal fluid a dilatation of the spinal cord directly above its narrowing appears. This theory is supported by the works by Josepshon et al. who produced a narrowing within the vertebral canal in rats (17). They used a knot pressing the dural sac, preventing the cerebrospinal fluid flow. At the next stage of research the MRI examination was performed. Cysts appearing within the spinal cord were located directly above and immediately below the narrowing.

A different situation can be observed in the case of CM, where the subarachnoid space becomes narrower but the cerebrospinal flow is possible (Fig. 3). Next, the factor triggering the development of SM is the so called Venturi effect (11, 51). It results from Bernoullie' law which says that during the fluid flow the sum of statistic and dynamic pressures is constant along each line of the flow. This means that when in a narrowed canal the fluid flow velocity increases, its pressure simultaneously decreases. Thus the Venturi effect increases the pressure gradient between the spinal cord and the subarachnoid space causing its dilatation with each systolic wave. In literature such a situation is described as the "suction effect" (11, 37). It was established that the syringomyelic cavites are not filled with the cerebrospinal fluid as was commonly thought. Their development takes place mainly as a result of

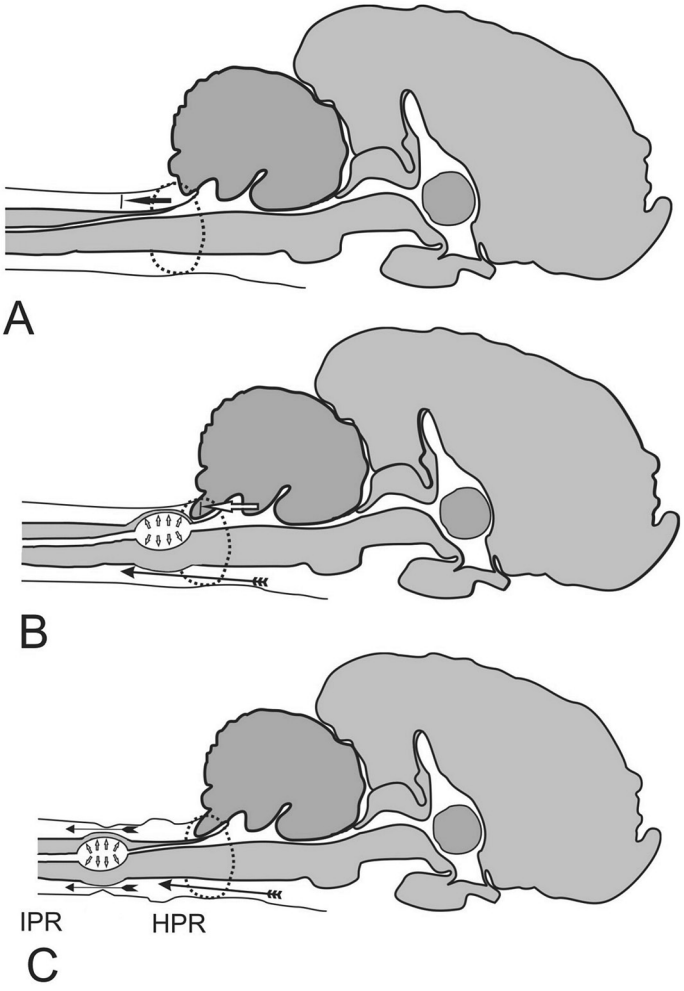

Fig. 3. Schematic illustration of syringomyelia development
A – Physiological condition. Dotted line illustrates foramen magnum. Black arrow indicates the direction of CSF flow.
B – Chiari malformation. Part of cerebellum is herniated through the foramen magnum (white arrow) disturbing the CSF flow at that level. Ventral CSF flow (black arrow) causes the Venturi effect which distends the spinal cord (suction effect) leading to syrinx formation just below the herniated part of cerebellum.
C – Chiari malformation. Narrowing of the subarachnoid space caused by cervical intumescense. HPR represents high pressure compartment whereas IPR shows intermediate pressure compartment. Increased CSF pulse pressure created by the cerebellum motion during cardiac cycle and functional obstruction of the subarachnoid space leads to Venturi effect occurrence at this level. Increased CSF velocity at the narrowing decreases CSF pressure and distends the spinal cord. Illustration shows why syrinxes are often located at or just below physiological or pathological narrowing of subarachnoid space.

accumulation of the extracellular fluid originating from the microcirculation in the spinal cord (11, 20, 22). The measurements of pressure directly inside the syrinxes permitted the statement that it is higher than in the subarachnoid space (22). As a result of the performed investigations the earlier theories were rejected. They assumed that the pressure in the syringomyelic cavites was lower than the pressure in the subarachnoid space. This would mean that the cerebrospinal fluid was forced into spinal canal against the gradient of pressures. It is commonly known that the pressure in the microcirculation of a given organ is always higher than in its parenchyma and thus the cavities appearing in the spinal cord are filled with the extracellular fluid. The presented assumption is a logical explanation of the SM pathophysiology.

Schmidt et al. described the mechanism which additionally disturbs the cerebrospinal fluid flow (44). They observed that in CKCSs with SM the jugular foramen is of a smaller size. It contains vessels carrying blood from the brain. In the case of its narrowing haemostasis takes place in the vessels which leads to the increase of intracranial pressure. Consequently there is an increase of the systolic pressure of the cerebrospinal fluid which probably is the driving force of the SM development. The authors stress that these are initial results and further micrometric investigations of the bone structures of the base of the skull are needed together with the description of the venous blood flow within the skull.

Most of the research concerning syringomyelia was performed using magnetic resonance imaging. Hu et al. (2011) described histopathological observations of CKCS spinal cord. In the cited work authors conducted microscopic examination of the spinal cord of two groups of dogs – CKCS with detected syringomyelia (with MRI usage) and individuals euthanized because of other diseases. Of the 14 isolated spinal cord specimens 12 revealed the presence of cavities within the spinal cord parenchyma (only 6 of which showed symptoms of the disease during life). In all cases there was a disruption of the ependyma lining in the central canal. Neurodegeneration and neuronal necrosis could be observed adjacent to the syrinx formation. In close proximity to cavities of symptomatic dogs there was presence of a large number of small blood vessels or condensed collagen tissue. A prominent feature of those vessels was the thickening of their walls which may suggest higher pressure (maybe as a compensatory mechanism to higher intra-cord pressure). Commonly there were also glial scars (proliferative astrocytes). The authors suggest that the presence of the latter may be responsible for the occurrence of clinical symptoms. In asymptomatic dogs in spinal cord parenchyma mainly oedema was present. In the future, microscopic examination may contribute to the understanding of pathophysiology of syringomyelia but it requires further research.

The breed particularly susceptible to CM development is the CKCS. However, it should be stressed that this disease was also diagnosed in King Charles Spaniels, Brussels Griffons, Yorkshire Terriers, Malteses, Chihuahuas, Miniature Dachshunds, Miniature/Toy Poodles, Bichon Frise, Pugs, Shih Tzus, Pomeranians, Staffordshire Bull Terriers, Boston Terriers, French Bulldogs, Pekingeses and a Miniature Pinschers. Despite a growing interest in CM, a detailed pathophysiology of the disease is not fully explained yet.

It is known that dogs representing the brachycephalic type as well as those belonging to the toy type breeds are highly at risk of that disease. Together with the CM diagnosis in a particular dog, the chance of the SM development clearly increases. That dependence results from the disturbances in the cerebrospinal fluid flow in dogs with CM which is a direct cause of SM. The described disease entities appear together so often that the CM/SM acronym is commonly used.

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