Syringomyelia
Information Sheet
What is it?

Syringomyelia is the term used to describe abnormal fluid filled cavities within the spinal cord, outside the central canal, that may or may not communicate with the central canal. These cavities are named syrinxes or syringes. Hydromyelia is the term used to describe fluid dilation of the central canal. The majority of syringomyelia cases diagnosed in dogs is associated to Chiari-like malformation. However, any condition that obstruct the flow of cerebrospinal fluid (CSF), such as congenital spinal cord and/or brain malformations, inflammatory and neoplastic diseases, may contribute to syrinx formation.

Affected Breeds

As seen in Chiari-like malformation, prevalence of syringomyelia has been commonly reported in brachycephalic toy breed dogs, in particular CKCS, King Charles spaniels, Griffon Bruxellois, Affenpinschers, Yorkshire terriers, Maltese, Chihuahuas, Pomeranians, Boston terriers, French Bulldogs and Papillons.

Clinical Signs

The most consistent clinical sign is neuropathic pain. Dogs with wider asymmetrical syrinx are more likely to be symptomatic. Other clinical signs include scoliosis (abnormal curvature of the vertebral column), especially in the cervical region (i.e. torticollis) and persistent scratching at the neck and shoulder area.
Chiari-like Malformation

Chiari-like Malformation Complex abnormality characterised by multiple anatomic malformations that create a mismatch of size between the brain and the skull. The skull is typically too small pushing part of the brain out through the opening at its base, with secondary crowding the spinal cord, obstruction of CSF flow and syringomyelia formation.

How does it occur?

The CSF is produced in the ventricles of the brain and circulates through the ventricular system and the subarachnoid spaces of the brain and spine cord.

Considering that the skull is a rigid closed space and that the circulation of CSF is greatly influenced by the pulsation of arteries associated with the heartbeat, the flow of the CSF must be in synchrony with the heartbeat. Disturbance of the normal free flow of CSF through the foramen magnum (large opening in the base of the skull) appears to be a major factor responsible for the formation of a syrinx in the cervical spinal cord. Other factors may, however, contribute to its formation.

When the normal pulsating CSF circulation is obstructed (e.g. cerebellar herniation), the smooth flow becomes a higher-pressure pulsatile flow and turbulence results in the spinal subarachnoid space (SAS), forcing CSF, vascular fluid or both into the parenchyma.

Another theory is that an obstruction to the CSF flow in the SAS results in a mismatch in timing between the arterial pulse peak pressure and CSF pulse peak pressure. An earlier arrival of peak CSF pressure compared to spinal arterial peak pressure encourages flow of CSF into the perivascular space.

The perivascular space changes in size during the cardiac cycle and is widest when the spinal arteriole pressure is low. If at that time peak CSF pressure is high then the perivascular space could act as a ‘leaky’ one-way valve. From the perivascular space, fluid flows into the central canal ultimately resulting in a syrinx (see MRI image below - the yellow arrows represent syringomyelia). A satisfactory explanation of how syringomyelia develops remains, however, unclear.

How to diagnose it?

Magnetic resonance imaging (MRI) is the goal standard to diagnose syringomyelia, since it also helps identifying other concomitant malformations/lesions of the brain and spinal cord.

Prognosis

Clinical signs are often progressive. However, in the majority of the cases, despite the progression, the patient’s quality of life does not seem to be severely compromised. Long term pain relief is required.

Treatment

Medical treatment:
- Drugs that reduce the CSF production (acetazolamide, cimetidine, omeprazole or furosemide);
- Analgesics (gabapentin, pregabalin, opioids, amantadine, etc).
- Corticosteroids (may provide pain relief although long-term therapy may be limited due to its side effects).

Surgical treatment:

There are no guidelines as to when surgery is indicated over medical management. It is overall agreed that surgery is advised in severe cases with poor response to medical treatment.

- Craniocervical decompression (re-establishing CSF flow). Recurrence of the clinical signs may, however, occur.