

# September 2014

## COMS / Syringohydromyelia screening

*NeuroNews* is an informal, but hopefully informative, newsletter covering a range of clinically relevant neurological topics. Cases detailed in these articles are patients kindly referred to me by veterinarians in South Australia and western Victoria/NSW.

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In recent years, the profession has become all too aware of the debilitating disease that is Caudal Occipital Malformation / Syringohydromyelia (COMS/SHM) syndrome. There is a well-known breed predisposition: the condition is prevalent in the Cavalier King Charles Spaniel, however many other breeds and types may be affected, including the Yorkshire Terrier, Toy Poodle, Pomeranian, Pug, Chihuahua and Brussels Griffon. With respect to the Cavalier King Charles Spaniel, it has been stated that 93% of the top stud dogs in the UK are closely related to one or more dogs diagnosed with the condition.

### PATHOGENESIS

The condition is thought to arise as a result of congenital dysplasia of the occipital bone, leading to increased pressure within the caudal fossa and interference with the dynamics of cerebrospinal fluid flow, with progressive fluid distension of the central canal of the spinal cord and/or development of fluid accumulations within the spinal cord parenchyma. The cervical cord is most commonly affected, but other segments may be involved.

### PRESENTATION

Signs may appear at any age, but typically between six months and three years. Patients most commonly exhibit signs consistent with cervical pain, although the degree is highly variable. Aberrant scratching behaviour, seemingly targeted around the caudal aspect of the skull, neck and forequarters is common; however contact with the head or body may not be made in such cases. Vocalisation during these episodes is not unusual. Scoliosis is an occasional finding.

Neurological findings are variable: patients may appear neurologically normal or may exhibit a range of signs, most commonly ataxia and/or paresis. In my experience, subtle signs of cerebellar dysfunction are occasionally noted, such as reduced menace response (the normal cerebellum is facilitatory to this response). Less commonly, "central cord syndrome" may be defined, in which the expanding syrinx leads to the development of progressive lower motor neurone signs in the forelimbs.

### DIAGNOSTICS

Although the classical presentation in a suspect breed allows a high index of suspicion, definitive diagnosis necessitates MR imaging of the post-tentorial skull, brain and cervical spine.

In addition to the presence of central canal dilation/syrinx, images of affected patients typically show a variable degree of occipital hypoplasia, cerebellar indentation/herniation and brainstem kinking (see Figs. 1 & 2 below). Lateral ventricles may also appear large, but this finding may or may not be related to COMS/SHM per se.

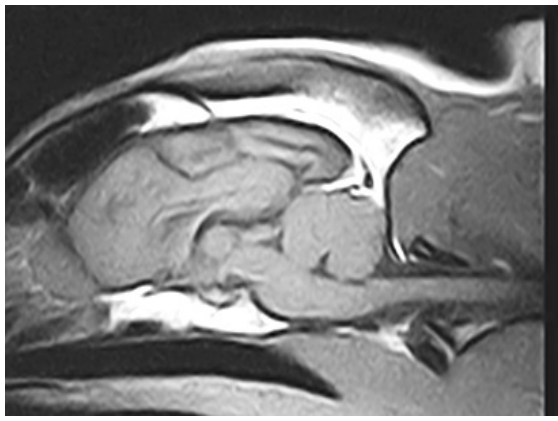


Fig. 1

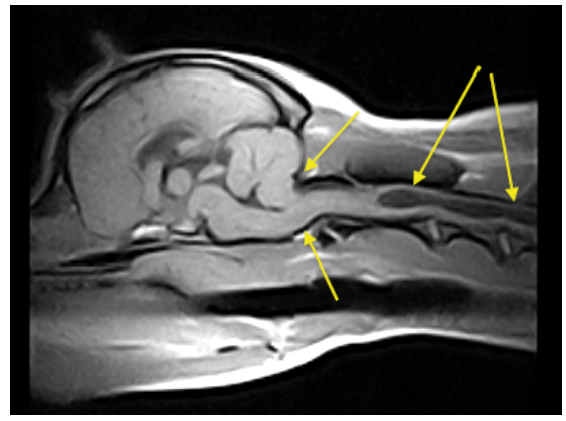


Fig. 2

Fig. 1: Normal canine neurocranial MRI (T1 sagittal)

Fig. 2: 3 y.o. FS Cavalier King Charles Spaniel referred to *NeuroVet*, having been exhibiting aberrant scratching behaviour for the previous 12 months: despite the extensive nature of the SHM, there was no evidence of ataxia or paresis (T1 sagittal)

#### ANCILLARY TESTS

Although CSF may show abnormality, findings are non-specific (increased protein and mild pleocytosis); moreover, the collection of CSF in such patients, especially via cisternal centesis, is discouraged, given the increased risks of morbidity and mortality.

#### TREATMENT

Medical management in my hands involves the administration of a variable combination of corticosteroids, proton pump inhibitors (in an attempt to reduce the rate of CSF production), diuretics and analgesics (most commonly gabapentin or pregabalin).

Surgery tends to be reserved for refractory cases. Various techniques have been described, typically involving partial dorsal laminectomy of the atlas and occipitoplasty. Results are encouraging, but variable. In a recent study, 81% of dogs showed some clinical improvement, but 25% of these dogs exhibited subsequent relapse.

#### PROGNOSIS

A fair longterm prognosis applies with appropriate treatment; however, due to financial constraints, many owners elect euthanasia rather than surgical management.

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#### SCREENING

**As reduction in the incidence of this distressing condition can only be achieved by exclusion of “affected” individuals from the breeding pool, in many countries, MR imaging is used as a screening tool, to assess suitability for breeding. *NeuroVet* is pleased to be able to offer this service to breeders in South Australia, Western Victoria and NSW and the Northern Territory.**

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I would be delighted to receive suggestions on topics to be covered in future editions of *NeuroNews*.

Best wishes,

Ian Douglas

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