

International Conference on Syringomyelia
Saturday 11th November 2006 – Royal Veterinary College, London. UK.

Report from the CM/SM Working Group Round table

Organiser – Rodolfo Cappello

Chairman Round Table– Clare Rusbridge

This meeting was hosted by the Cavalier Club UK and sponsored by Boehringer Ingelheim Ltd and enabled veterinarians with an interest in syringomyelia to share views and ideas on this poorly understood disease. In addition to inviting veterinarians from Europe, Canada and the USA, the Cavalier Club UK also included breed club representatives from across the UK with the aim of disseminating information about this disease to the wider breeder and dog owning population.

The round table meeting took place after very interesting and stimulating oral presentations covering a wide range of subjects including pathogenesis of syringomyelia associated pain, surgical management, MRI studies, new technological advances, experiences from France and update on progress of the CKCS genome scan.

Members present

Rodolfo Cappello (Royal Veterinary College), Clare Rusbridge (Stone Lion Veterinary Centre), Harvey Carruthers (Stone Lion Veterinary Centre), Laurent Cauzinille (Centre Hospitalier Vétérinaire Fregis), Nick Jeffery (Cambridge Veterinary School), Catherine Louglin (Long Island Veterinary Specialists), Dominic Marino (Long Island Veterinary Specialists), James Anderson (Glasgow Veterinary School) Martin Deutschland (Chestergates Referral), Imelda McGonnell (Royal Veterinary College) and Steven Dean (Dogs World Magazine)

The meeting was conducted in the presence of representatives from the UK CKCS club; the UK Kennel Club (Jeff Sampson) and UK, Eire, French and North American CKCS dog owners.

The Round table meeting had, and achieved, the following objectives

1) Agreement for a name for the canine disorder

The participants recognised that there are uncertainties about the correct terminology to use. The pros and cons of the terms syringomyelia, syringohydromyelia, hindbrain herniation/ descent, Chiari malformation, occipital hypoplasia and caudal occipital malformation syndrome were discussed and the main points for and against each term are summarised.

Syringomyelia - generally this term is accepted however it was rejected as a name for the condition that predominantly occurs in Cavalier King Charles spaniels and other small breeds because there are many potential causes of syringomyelia.

Hindbrain herniation / descent – this term was rejected because hindbrain is an embryological term and does not describe the adult anatomy. Equally, the cerebellum is only a component of the hindbrain.

Chiari malformation – There was general resistance to use this term because it uses the name of the first scientist that described the disease. However in humans this term no longer reflects the original description of the disease but any condition

characterised by reduced posterior fossa volume and caudal descent of the brain stem and cerebellum. It was pointed out that although there is general resistance to the use of “proper nouns” to describe diseases in veterinary medicine it is not without precedent especially where the name is a simple term used to describe a complicated process e.g. “Wallerian degeneration”.

Occipital hypoplasia and Caudal occipital Malformation Syndrome (COMS) -

These terms were rejected because there is no proof yet that the condition is related to either a malformed or hypoplastic occipital bone(s). Current evidence suggests that there may be other significant factors in the pathogenesis. In addition these terms can be confusing as for example the term COMS may imply the malformation only or the malformation and syringomyelia.

The majority vote was for the term **Chiari-like malformation and syringomyelia (CM/SM)** to be adopted (at the current time). This term was perceived to have the following advantages.

1. Chiari malformation is accepted for the description of the disease in the human species and is the most commonly used term in scientific publications.
2. Chiari-like malformation refers to the complex syndrome seen in the human species however the “like” implies some differences in the canine.
3. The term can be easily abbreviated to CM/SM– having a simple acronym is especially important to dog breeders and owners.
4. The term Chiari-like malformation (CM) can be used to distinguish dogs that do not have syringomyelia

Chiari-like malformation (CM) is currently defined as decreased caudal fossa volume with caudal descent of the cerebellum, and often the brainstem, into or through the foramen magnum.

Syringomyelia (SM) is currently defined as a condition that results in the development of fluid-containing cavities within the parenchyma of the spinal cord as a consequence of abnormal cerebrospinal fluid movement.

2) At the request of the UK CKCS club formulate and agree on breeding guidelines for CM/SM.

Clare Rusbridge presented the very early but promising results of the breeding program in the Netherlands. It was suggested that before genetic studies are completed that “commonsense” strategies aiming to limit possible widespread dissemination of the disease be implemented. The main aim was to limit early onset and potentially painful SM and to avoid using such dogs in a breeding program. The current breeding guidelines were discussed and were simplified and modified (**Appendix 1**) The presence or absence of the Chiari-like malformation (CM) was dropped from breeding guidelines because of 1) of the ubiquity of this malformation within the CKCS population 2) Lack of uniformity between veterinarians at recognising and consistently grading the severity of CM 3) the lack of evidence that apparent severity of CM was related to severity of syringomyelia. It was agreed that MRI screening of subsequent generations should be continued so that these early breeding guidelines could be adapted as more information on the heritability becomes available.

3) Formulate and agree on a pain scoring system that could be used in prospective studies

Future prospective studies require uniformity in grading the severity of the clinical signs. An existing pilot system was adapted (*Appendix 2*).

4) Implement protocols for dealing with pathological material.

One of the recurring themes during the oral session was that the pathogenesis of CM/SM is not understood and to improve understanding of the disease there is a need for post mortem studies at different stages of the disease and ages of dog. All cavalier King Charles spaniels that die for related or unrelated causes would be valuable and the following centres are willing to participate in this program. Breeders and owners willing to donate their pet for this purpose should contact:

- Professor Nick Jeffrey, Department of Veterinary Medicine, University of Cambridge, Madingley Road, Cambridge, CB3 0ES tel 01223 337621 ndj1000@cam.ac.uk
- Dr Jim Anderson, Glasgow University Gvsa07@udcf.gla.ac.uk
- Dr Rodolfo Cappello The Royal Veterinary College, University of London RCappello@RVC.AC.UK
- Dr Curtis Dewey, Cornell University
- **Fetal and neonatal specimens**

Another area of great interest is developmental studies looking at anomalies in different stages from the early growth in the uterus to the maturity of the dog. These studies would be also useful to confirm or characterize the genetic defect for CM/SM. This area of research is difficult because of a requirement for foetuses and young puppies therefore a request was also made for any aborted foetus or puppies that die for related or unrelated disease. Any dog owner or veterinarian with such material should contact Dr Imelda McGonnell (Dept. Veterinary Basic Sciences, Royal Veterinary College, Royal College St, London, NW1 0TU). Tel 020 7468 1223 Imcgonnell@RVC.AC.UK

NOTE - As the nervous system degenerates rapidly and must be handled appropriately contact with these centres should be made as soon as or ideally before the pet has been euthanatized.