

Report from the Chiari-Like Malformation and Syringomyelia Working Group Round Table

Organizer RODOLFO CAPPELLO and Chairman Round Table CLARE RUSBRIDGE

THIS MEETING was hosted by the Cavalier Club UK and enabled veterinarians with an interest in syringomyelia (SM) to share views and ideas on this poorly understood disease. In addition to inviting veterinarians from Europe, Canada and the United States, the Cavalier Club UK also included breed club representatives from across the United Kingdom with the aim of disseminating information about this disease to the wider breeder and dog owning population.

The round table meeting occurred after oral presentations covering a wide range of subjects including pathogenesis of SM associated pain, surgical management, MRI studies, new technological advances, experiences from France and update on progress of the Cavalier King Charles Spaniel (CKCS) genome scan.

Members: Rodolfo Cappello (Royal Veterinary College), Clare Rusbridge (Stone Lion Veterinary Centre), Harvey Carruthers (Stone Lion Veterinary Centre), Laurent Cauzinille (Centre Hôpitalier 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 Today 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.

OBJECTIVES

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

Agreement for a Name for the Canine Disorder

The participants recognized that there are uncertainties about the correct terminology to use. The pros and cons of

the terms SM, syringohydromyelia, hindbrain herniation/descent, Chiari malformation, occipital hypoplasia, and caudal occipital malformation syndrome (COMS) were discussed and the main points for and against each term are summarized.

SM. SM generally this term is accepted, however, it was rejected as a name for the condition that predominantly occurs in CKCS and other small breeds because there are many potential causes of SM.

Hindbrain Herniation/Descent. 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. Chiari malformation there was general resistance to use of 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 characterized 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 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 SM.

CM/SM

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. CM 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 CM can be used to distinguish dogs that do not have SM.

CM. 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.

SM. 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.

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 A) The presence or absence of the CM was dropped from breeding guidelines because of (1) the ubiquity of this malformation within the CKCS population, (2) the lack of uniformity between veterinarians at recognizing and consistently grading the severity of CM, (3) the lack of evidence that apparent severity of CM was related to severity of SM. 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.

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 B).

Implement Protocols for Dealing with Pathologic 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 postmortem studies at different stages of the disease and ages of dog. All CKCS that die for related or unrelated causes would be valuable and the following centers 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 fetuses and young puppies therefore a request was also made for any aborted fetus or puppies that die for related or unrelated disease. Any dog owner or veterinarian with such material should contact Dr. Imelda McGonnell (Department of Veterinary Basic Sciences, Royal Veterinary College, Royal College St., London NW1 0TU, UK). Tel 020 7468 1223 Imcgonnell@RVC.AC.UK

Note—as the nervous system degenerates rapidly and must be handled appropriately contact with these centers should be made as soon as or ideally before the pet has been euthanized.

Appendix A

Revised CKCS MRI Screening and Breeding Recommendations

These breeding recommendations are made using current information and in response to CKCS breeder request for guidelines. It has yet to be proven if this guide is appropriate. The aim of these recommendations is to reduce the incidence of symptomatic SM in the breed, not to create litters of puppies guaranteed not to have SM as the chance of producing an affected dog cannot be predicted without knowing the inheritance.

Notes

The age cut off at 2.5 years has been decided so as to tie in with MVD recommendations and because most dogs with symptomatic SM will show signs before 3 years of age.

The following categories from the previous guidelines have been removed because of difficulty in accurately interpreting

Previously A* – now A

Previously B – now C

It is recommended

- (1) That both the sire and the dam of a proposed mating are screened (any unscreened dog should be assumed to be “D”).
- (2) Offspring of any mating should also be MRI screened before breeding.

- (3) Any dog screened before 2.5 years old has a second screen when older.
- (4) That dogs are screened from 6 months of age.
- (5) That if a limited (“mini”) MRI screen is performed that
 - (a) the minimum area covered is from the level of the interthalamic adhesion to cervical vertebrae 5 (C5);
 - (b) both TW1 and TW2 sagittal images are obtained in addition to TW1 and /or TW2 transverse images through the upper cervical spinal cord; and
 - (c) an assessment is also made for presence/absence of ear disease and ventricular enlargement.
- (6) That interpretation of images is made by Diplomate level radiologists, neurologists and, in special circumstances, by surgeons with recognized expertise in this area.

Grade	Age (Years)	Syringomyelia	Breed to
A	Over 2.5	Absent or <2mm central canal dilatation in the C2-C4 region only	A, C, D
C	Under 2.5	Absent	A Re scan after 2.5years
D	Over 2.5	Present Asymptomatic	A
E	Under 2.5	Present Asymptomatic	Do not breed
F	Any	Present Symptomatic	Do not breed

Appendix B

CM/SM Pain Score and Clinical Signs

Pedigree Name: _____

Registration number _____ Microchip number _____

Date of birth: _____ Call name _____ Owner's name _____

Colour B B/T R T Gender M MN F FN Weight

Pain Score	Frequency Vocalization	Frequency Scratching	Exercise Ability
0	None	None	Normal
1	< 1/week	< 1/day	Normal
2	1/week	≥ 1/day	Normal
3	> 1/week	> 1/day	Normal
4	> 1/week	> 1/day	Activity compromised

Dogs scored according to the most severe clinical sign for example a dog vocalising once daily but shoulder scratching less frequently would be scored 3.

Pain score No pain or neurological dysfunction

Possible Signs of Pain/Neurological Dysfunction

Signs	Frequency	Age of onset	Signs	Frequency	Age of onset
Shoulder scratching (indicate side)			Scoliosis	N/A	
Scratching elsewhere (indicate site)			Thoracic limb ataxia	N/A	
Rubbing ears			Thoracic limb weakness	N/A	
Rubbing mouth			Thoracic limb lameness		
Cervical pain			Pelvic limb ataxia	N/A	
Thoracic pain			Pelvic limb weakness	N/A	
Lumbar pain			Pelvic limb lameness		
Screaming when scratching			Vestibular dysfunction (indicate side)	N/A	
Screaming when excited			Facial nerve dysfunction (indicate side)	N/A	
Screaming when touched			Seizures		
Screaming when change head position			Fly catching		
Screaming when jumping			Collapse during exercise		
Screaming for no apparent reason			Cramping during exercise		

N/A, not applicable.