

## Dr. Rusbridge, Veterinarian & Chiari Researcher

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Dr. Clare Rusbridge is a veterinary neurologist working at the Stone Lion veterinary Centre, which is a specialist referral practice in Wimbledon, London. For several years she has studied Chiari and syringomyelia in a breed of dog known as Cavalier King Charles Spaniels (CKCS) and has published multiple papers on the subject.



Is the canine form of CM/SM similar to the human form? How is it diagnosed and treated? What can we learn from man's best friend?

To find out, we put Dr. Rusbridge In The Spotlight...

### How did you come to the point of studying and treating Cavalier King Charles Spaniel with Chiari and syringomyelia?

**R:** In 1997 I was treating a dog called Beau for weakness of a fore limb. He had a tendency to scratch at his shoulder area when he walked. The action was quite unlike anything I had seen as the dog tended to bicycle one of his back legs in the air as he walked. Wearing a neck collar, movement and excitement seemed to be the trigger. This behaviour is a probably a reflection of allodynia and is very common with CKCS with syringomyelia but at the time I couldn't understand it and resolved to get an answer. At the time, spinal MRI for animals was not available so finding the answer took a few years by which time there were a few other cases (all CKCS). When Beau and 2 other CKCS were confirmed to have syringomyelia secondary to a Chiari malformation I wondered if there may be a tendency for this in the breed. I wrote a letter describing the scratching signs to the most popular UK veterinary journal. The letter was published on the Saturday and on the Monday morning I have 30 telephone calls from vets across the country all with similar cases. From that point I have been researching the genetics, the pathogenesis (why some dogs get syringomyelia and/or pain and some don't) and treatment. I am now close to completing a PhD thesis on the subject.

### I've noticed you tend to not use the term Chiari when referring to the condition in dogs, is there something behind this?

**R:** It is an attempt to be scientifically correct. It is typical in (human) medicine to call a condition after the person who discovered it but in veterinary medicine it is more usual to use a scientific description. Also the dog condition is not the same as the human definition. The type 1 malformation was originally described by Hans von Chiari as "elongation of the cerebellar tonsils and the medial part of the inferior cerebellar lobes into cone-like projections, which accompany the medulla into the spinal canal" (translated). Dogs do not have cerebellar tonsils and it is not always necessary for a CKCS with syringomyelia to have a significant cerebellar herniation. The fundamental primary problem is that the posterior fossa is small and the foremen magnum is overcrowded. However in recent years in (human) medicine the term Chiari malformation has become shorthand for a wide range of abnormalities not necessarily consistent with Hans von Chiari's original description type 1 malformation, but all characterized by decreased posterior fossa volume with caudal descent of the cerebellar tonsils. This is much more in keeping with the dog condition so I am leaning towards calling it Chiari malformation especially as it is a simpler term for owners to remember.

### What leads you to suspect occipital hypoplasia (Chiari) in a dog you are evaluating?

**R:** PAIN !!!!

The most common signs are unexplained sensitivity especially in the head and neck area. Other signs of syringomyelia are similar to the human condition e.g. weakness, scoliosis. However many are asymptomatic, most CKCS have a Chiari malformation and ~ 60% have syringomyelia.

### Do you always perform an MRI to confirm the diagnosis?

**R:** The diagnosis can only be confirmed with a MRI scan. However MR imaging is expensive and therefore not always an option and in some dogs the condition is suspected but not confirmed.

### Is pet imaging (as they say in the US) readily available in the UK?

**R:** It is readily available - there are many private and university hospitals that have on or off site MRI facilities, in the UK most people are no more than 3 hours away from a facility and it is usually possible to have an appointment within 2 weeks. It is expensive. The running costs are no different to a human facility and the dog has to be anaesthetised which increases the price. In the UK pet health insurance is increasingly popular and most policies will cover the cost of an MRI work up. In my practice the Stone Lion Veterinary Centre we are very fortunate to have an on site MRI scanner. In addition many veterinary centres with MRI facilities offer a low cost scanning service for CKCS breeders so that potential breeding stock can be screened economically.

### In people, the surgical decision can be very difficult; what are your criteria for recommending surgery for an affected dog?

**R:** For canine patients, surgical management is indicated when drugs do not control discomfort or when neurological deficits are present. Medical management may be chosen for patients with only mild pain, when finances do not allow surgical management, or when surgical management has failed to resolve the signs.

### Is the surgery essentially the same? Do you open the dura?

**R:** I perform a suboccipital decompression then a partial C1 laminectomy. Yes I open the dura.

### How is occipital hypoplasia in CKCS different from Chiari in people?

**R:** The main difference is that there is not always a great cerebellar herniation.

**What percent of CKCS have, or are likely to develop, the condition?**

**R:** Most CKCS seem to have a small posterior fossa and about 60% have syringomyelia. It is difficult to say what proportion of the dogs have clinical signs because the long term studies of young asymptomatic dogs with syringomyelia have not yet been completed and my hospital population is naturally skewed towards affected dogs.

**You have been studying the CKCS for some time now, what have you found?**

**R:** It appears to be inherited but the inheritance is complex and may involve more than one gene. A breed genome scan is now underway. There is still a great deal more to find out and explain. We are currently trying to determine if there are other factors that influence the development of syringomyelia with Chiari malformation; the precise nature of the bony abnormality and complete long term studies on treatment. I wish to state that I do not experiment on these dogs nor keep them in a laboratory. I treat pets so my research is clinical or involves DNA.

**Do you discuss your findings/experiences with neurosurgeons in the UK or US?**

**R:** The Stone Lion Veterinary Centre is close to the Atkinson Morley hospital in London, sadly now closed. This hospital was entirely neurology / neurosurgery and I was very fortunate to be able to share my experiences with the surgeons and radiologists there. My original paper was written with a human neuroradiologist and I was also able to watch the surgery performed by Mr Henry Marsh (as of the television program "your life in his hands" fame). I have attended several "human" meetings most notably a neurosurgery conference in Philadelphia and ASAP forum in New York. Many experts in the Chiari field were present and I was able to learn from their experience. In particular neurosurgeons Dr Milhorat and Dr Iskandar have been most helpful and interested. A veterinary review paper is to be published shortly and 2 "human" experts Dr Iskandar and Dr Griez are co-authors with me. However by far the most valuable learning experience was speaking to people afflicted with syringomyelia. As dogs cannot express what they feel, human sufferers were able to, and still give me, a greater insight into the condition. I am particularly interested in factors that affect pain and it would be very interesting to do a cross species comparison, e.g. many dogs seem to be worse with certain weather conditions and I am given to understand that the same may be true for humans.

**In the human medical literature, study after study has found little to no correlation between the amount of tonsillar herniation and symptoms or clinical outcome. Do you think the definition (and focus) of the condition needs to change?**

**R:** Absolutely yes!!! I think the focus should be on whether foramen magnum and cerebrospinal fluid flow is obstructed, not the size of the herniation

**Since you use the term occipital hypoplasia (underdevelopment), do you believe the core problem is a skull that is too small? Or, is it more complex than just size?**

**R:** Yes the back of the skull is too small but I also think there may be other contributory factors and this is one of main focuses at the moment.

**You're now working on compiling a DNA database. How are you going about this and what do you hope to accomplish?**

**R:** The data base, about 10000 dogs and 1000 DNA samples and still growing, was compiled and organised by my co-worker Penny Knowler. The genome scan is underway. We are working with Guy Rouleau and Zoha Kibar, prestigious neurogeneticists at Montreal University, who are ably helped by hard working research assistant Melanie Barnard. We hope to identify the genetic defect(s) and then the group plan to extend the study to humans

**In one of your updates posted on the web, I was struck by a comment that the entire breed tends to have a very small occipital skull region. Do you think it's possible the condition is not due to a genetic anomaly, but rather may fall within what might be considered normal for the breed?**

**R:** What is normal? - the answer is a wolf ! A dog breed is created by breeding closely related individuals together to create a "type" with little genetic variation. Therefore it is perfectly possible for many of them to have a genetic anomaly just as most CKCS have adorable cute faces and a friendly disposition. Most CKCS also develop heart disease, mitral valve disease, and many have a platelet abnormality.

**If you do identify a gene, or set of genes, from the database, is it likely that this finding would translate to human Chiari?**

**R:** I don't know - it is certainly a place to start

**A recent article in Nature (Matsuoka T et. al.) proposed that Chiari is due to a transcription error resulting in the clivus not forming properly. Do you have any thoughts on this?**

**R:** The findings are really interesting particularly in view of work we are currently completing.

**Have you seen anything similar to Chiari/syringomyelia in any other animals?**

**R:** Arnold Chiari syndrome, or Chiari type II, has been described in cattle and it is possible to induce Chiari in hamsters by overdosing the developing embryos with vitamin A.

**Are you surprised that the condition exists in both people and dogs, given that people walk upright and dogs don't?**

**R:** Not at all

**Do you think our understanding of these conditions, in both people and dogs, will evolve over the next 5-10 years?**

**R:** Definitely! The CKCS represents a unique opportunity for understanding Chiari malformation because it is a naturally occurring disease not a laboratory induced condition and therefore if we better understand the pathophysiology in the dog (i.e. how syringomyelia develops) then we may better understand it in humans. As these dogs are beloved pets receiving long term veterinary care we also have the opportunity to get better insight into treatment.

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If you would like more information about CM/SM in the CKCS, a DVD of a lecture by Dr. Rusbridge is available for purchase. (Note: Conquer Chiari and this publication have no affiliation with this DVD offer)

**Live Syringomyelia Seminar** given by Clare Rusbridge to the CKCS Club UK in December, 2005.

Cost \$20 US/Canadian, £10, or 15 Euros

Lecture Synopsis: Causes, History of the name, Clinical signs, Diagnosis, Treatment -medical and surgical, Future

For additional DVD with Breeders questions \$30, £15 or 20 Euros ( for both)

All profits go towards Syringomyelia DNA Research.

To order a copy contact Penny Knowler - [penny.knowler@ntlworld.com](mailto:penny.knowler@ntlworld.com) or Karlin Lillington [karlin@cavaliertalk.com](mailto:karlin@cavaliertalk.com)

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