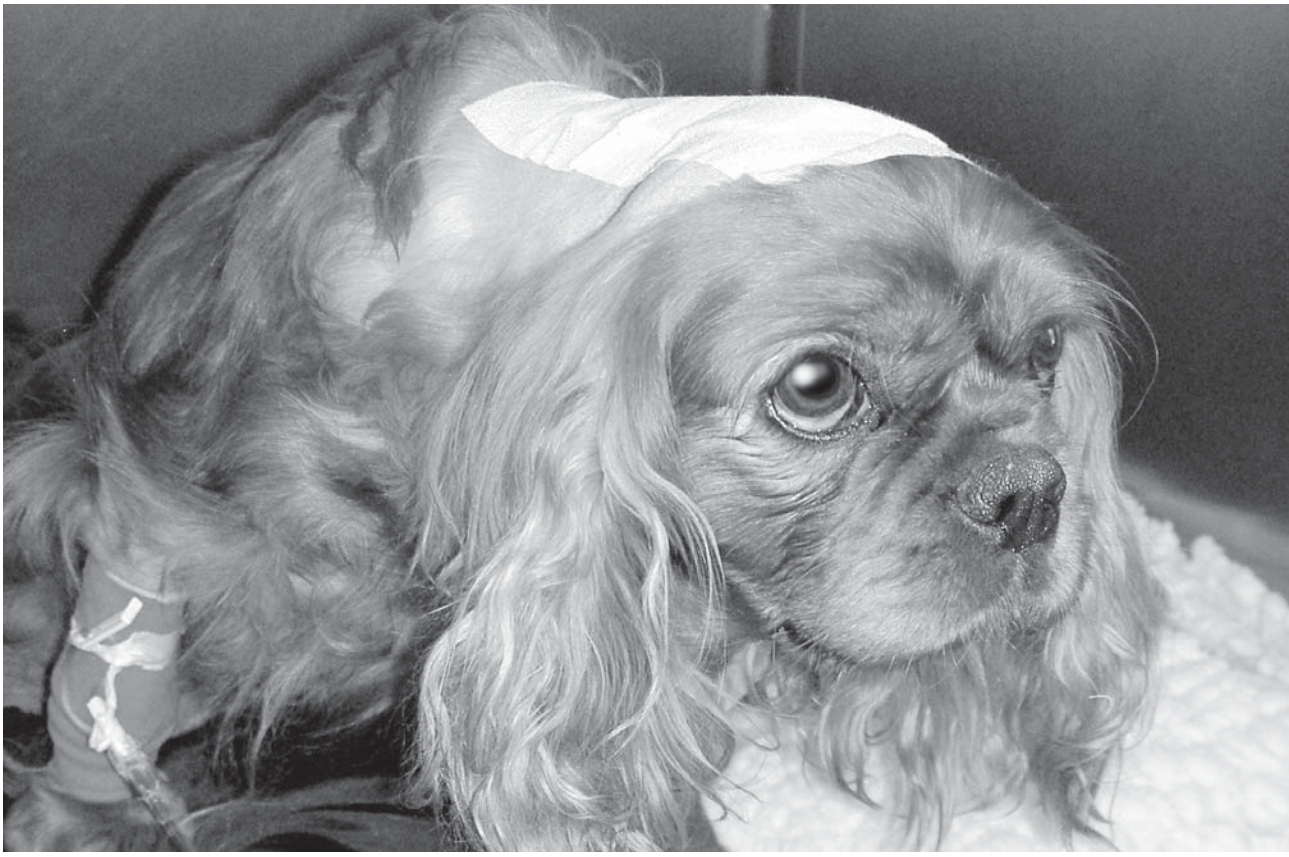

Section 6

Surgical management

Chapter 6.1

Chiari-like malformation with syringomyelia in the cavalier King Charles spaniel; long term follow up after surgical management

C. Rusbridge

Stone Lion Veterinary Centre, 41 High Street, Wimbledon Village, London, SW19 5AU

Accepted – Veterinary Surgery

Introduction

Syringomyelia is a condition that results in fluid-containing cavities within the parenchyma of the spinal cord as a consequence of obstruction to cerebrospinal fluid (CSF) movement. It is increasingly recognised in the dog and one of the most common causes is Chiari malformation - defined as decreased caudal fossa volume with caudal descent of the cerebellum, and often the brainstem, into the vertebral canal[1]. Chiari-like malformation (CM) is particularly common in the cavalier King Charles spaniel (CKCS)[2, 3, 4]. The condition is characterized by a mismatch between the caudal fossa volume and its contents, the cerebellum and brainstem, and consequently there is insufficient room for the neural structures, which are forced caudally, obstructing the foramen magnum and the pressure wave of CSF emanating from the head during arterial pulsations. The pathogenesis of syringomyelia is much debated[1, 5]. There is increasing agreement that the syrinx fluid is not CSF and is most likely extracellular fluid that coalesces within the central canal and/or spinal cord substance as a consequence of abnormal pressure differentials between the spinal cord and subarachnoid space[5, 6].

Pain is a predominant feature of the disease in humans and animals with ~ 80% (human) and 35% (dog) respectively reported to experience discomfort[7, 8]. Owners may describe signs suggesting head, spinal or other discomfort. The pain may be difficult to localise on clinical examination as it is often intermittent. Common historical features include yelping after sudden posture change or being more uncomfortable in the evening or early morning. A recent study found that syrinx width is a predictor of pain and suggested that spinal cord dorsal horn damage may result in a neuropathic pain syndrome[8]. Affected dogs may show behavioural signs suggestive of allodynia, *i.e.* pain arising in response to a non-noxious stimulus, for example they appear to dislike touch to certain areas of skin (ear, neck, forelimb or sternum). Dogs with a wide syrinx may also scratch, typically on one side only, while the dog is walking and often without making skin contact [1]. The behaviour is suggestive of dysesthesia - *i.e.* a spontaneous or evoked unpleasant abnormal sensation, described by some humans as an intense feeling of insects crawling on the skin or a painful burning itchiness[9].

Surgical therapy has been recommended[10] to improve the dog's quality of life and to retard clinical and radiographic progression of the syrinx. The most common procedure is cranial cervical decompression (also described as suboccipital decompression or foramen magnum decompression) in which most of the supraoccipital bone and dorsal laminae of the atlas are removed (with or without a durotomy) to decompress the foramen magnum[10,11,12]. In this study the case histories for 15 CKCS which had surgical management of Chiari-like malformation/syringomyelia (CM/SM) at least 12 months previously were reviewed retrospectively.

Material and methods

15 consecutive cases which had surgical management of CM/SM over a 6 year period and with at least 12 months follow-up were evaluated retrospectively (Table 1 and 2). Diagnosis was made on the basis of neurological examination and magnetic resonance imaging (MRI) of the brain and cervical spinal cord. The dogs also had haematology and biochemistry. CSF analysis was not performed in any case because of the perceived risk of doing this procedure in dogs with a cerebellar herniation and wide syringomyelia, because the brain and spinal MRI did not support a diagnosis of central nervous system inflammation and because CSF in cases with syringomyelia typically has an inflammatory pleocytosis [2] making interpretation difficult. Pain was determined by historical features such as yelping, scratching behaviour and/or apparent discomfort or avoidance of being touched over the ear, neck, shoulder or sternum together with clinical features such as spinal hyperaesthesia or abnormal head posture. The cases were assigned, retrospectively, a pre- operative pain score, as follows:

- 0 = No pain
- 1 = occasional (< 1 per week) signs of facial, ear (e.g. rubbing) or spinal pain and / or yelping
- 2 = Mild neuropathic pain - occasional shoulder scratching when excited, on waking and when touched.

Does not scream whilst scratching.

- 3 = – Moderate neuropathic pain - consistent shoulder scratching at exercise, when excited and when touched. May occasionally cry when scratching. May appear to dislike touch to certain body parts for example one ear. Owner considers a normal active dog in other respects.
- 4 = Severe Neuropathic pain - consistent shoulder scratching at exercise, when excited and when touched or at other times. Ears or other body parts cannot be touched and/or or groomed. Frequently screams. May adopt unusual body posture when sleeping. Owner feels dog's activity is compromised by the disease.

Other neurological signs e.g. scoliosis, long tract signs (ataxia and /or pelvic limb weakness), ventral horn signs (lower motor neuron signs thoracic limbs) and seizures were recorded. A decision was made for surgery based on severity of pain and lack of response to medical management (10 of 15 dogs) or because the owners were concerned that the dog's signs may be progressive (5 of 15 dogs) (Table 1).

A cranial cervical decompression was performed by the same surgeon in all cases. 30 minutes prior to surgery the dogs received a premedication of 100mg gabapentin (Neurontin Parke-Davis, Eastleigh, UK) per os (this drug was continued post-operatively starting 8 hours after the initial dose), 4mg/kg carprofen (Rimadyl, Pfizer Limited, Sandwich, UK) subcutaneously, 0.2mg/kg methadone hydrochloride (Physeptone, Martindale Pharmaceuticals, Romford, UK) intramuscularly, 0.02mg/kg acepromazine maleate (ACP, Vericore Novartis Animal Health, Royston, UK) intramuscularly. Anaesthesia was induced using 4mg/kg intravenous propofol (Rapinovet, Schering-Plough Animal Health, Welwyn garden City, UK) and maintained with isoflurane (1-3% in 100% oxygen). After induction, 20mg/kg Cephradine (Velosef, Bristol-Myers Squibb, Dublin, Ireland) was administered intravenously. A repeat dose was given every hour during surgery until 1 hour postoperatively (typically 3 doses including premedication dose). A pulse oximeter (Nonoin, Kruuse, A/S Denmark) and oesophageal stethoscope were used to monitor heart rate and adequacy of ventilation. Diastolic blood pressure was recorded using an ultrasonic Doppler flow detector and temperature using a rectal probe oesophageal thermometer. All dogs received intravenous fluids during surgery which was continued until 24 hours postoperatively. The dogs were positioned in sternal recumbency and the head was flexed over a vacuum support cushion (Buster vacu support, Kruuse, A/S Denmark). Tape was used to prevent movement (Fig 1).

Figure 1 CKCS positioned for cranial cervical decompression. The head was flexed over a vacuum support cushion and secured using tape.

A dorsal approach was made to the supraoccipital bone and atlas. The rhomboideus, splenius and occipital muscles were reflected aside and the supraoccipital bone and atlas was exposed. A supraoccipital craniectomy and partial C1 laminectomy was performed in all cases. The extent of the bone removal is depicted in figure 2.

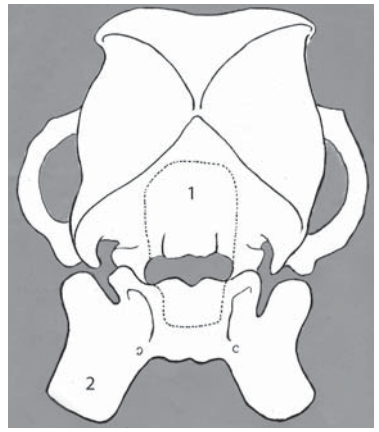

Figure 2 Cranial cervical decompression. The dotted line delineates the extent of bone removal from the supraoccipital bone and dorsal laminae of the atlas. 1. supraoccipital bone, 2 wing of atlas.

The bone was burred using a Hall Sugairtone 2 (Linivatec Corporation, Largo, Florida) until egg-shell thickness and then removed with rongeurs and dental instruments. Care was taken in the region of the vermiform impression as the bone was extremely thin and very little drilling was required. The atlantooccipital membrane was removed using a scalpel blade and bipolar electrocautery. Associated with the atlantooccipital membrane was a tight constricting band of connective tissue across the foramen magnum which was also removed. 8/0 Vicryl (Ethicon, Johnson and Johnson St Steven-Wolouwe Belgian) stay sutures were placed in the dura. Using the stay suture to identify and support the outer meninges (Fig 3), the dura and subarachnoid membranes were incised in a cross shape as depicted in figure 4.

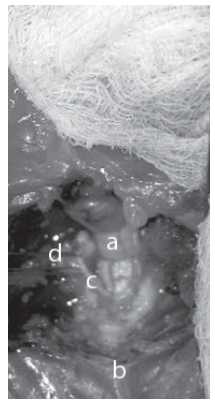

Figure 3 Cranial cervical decompression. The dura and arachnoid meninges have been incised and are supported by a stay suture. The spinal cord and the cerebellar vermis can be visualised though the meningeal defect. The dogs head is towards the top of the picture. a. caudally displaced vermis b. dorsal spine C2 c. dura and arachnoid meninges d. stay suture

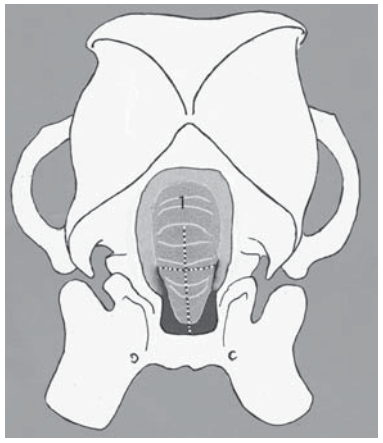

Figure 4 Cranial cervical decompression. The dotted line delineates the incision through the meninges. 1. cerebellar vermis.

This shape of incision was chosen because it allowed the best visual decompression of cerebellum and medulla. In some dogs bleeding occurred from the dural vessels which were controlled with bipolar electrocautery and / or micro-porous polysaccharide beads (Hemablock, Abbott Laboratories, North Chicago). The neural structures were inspected for adhesions which were removed if present. The C1 laminectomy was continued until ~ 3mm caudal to the tip of the cerebellar vermis; typically to the level of the attachment of the dorsal spinous process of C2. The resulting kite shaped defect in the meninges was either left open and covered with absorbable collagen sponge (Lysostypt Braun Aesculap, Rueeggisingerstr, Switzerland) (earlier cases listed in Table 1a) or expanded by patching with 2 layers of a biocompatible collagen matrix (Vet BioSIST, Cook/Global Veterinary Products, SurgiVet, Smiths Medical Pm inc N7 W22025 Johnson Road, Waukesha, WI USA 53186). If possible the patch was sewn to the dura using the Vicyl 8/0 stay sutures however in case 14 manipulation of the dura resulted in repeated excessive haemorrhage and so the patch was simply laid over the defect overlapping the dural edges by at least 1cm. To aid with haemostasis an absorbable collagen sponge (Lysostypt Braun Aesculap, Rueeggisingerstr, Switzerland) was then placed over the entire defect. The muscle layer was closed with 2/0 PDS, subcutaneous layer were closed with 2/0 Vicryl and the skin defect was closed with skin staples (Skin Mate Skin Stapler Animalcare Ltd Dunningtom York).

Postoperatively the dogs received 0.2mg/kg methadone (Physeptone, Martindale, Pharmaceuticals, Romford, UK) every 4 hours. When the dog had appeared consistently comfortable over an 8 hour period this was switched to 0.015mg/kg of Buprenorphine (Vetergesic, Alistoe Animal Health, Sheriff Hutton, UK) every 6 hours which then gradually withdrawn at a rate determined by the degree of pain behaviour displayed by the dog. Carprofen (Rimadyl, Pfizer Limited, Sandwich, UK) at 2-4mg/kg per day and gabapentin (Neurontin Parke-Davis, Eastleigh, UK) at 100mg every 8 to 24 hours were continued for at least 2 weeks post operatively then gradually withdrawn in all dogs with the exception of dog 12 who was continued on twice daily gabapentin and carprofen. All dogs also received 20-30mg/kg of Cefalexin (Ceporex 250mg; Schering-Plough Animal Health, Uxbridge, UK) twice daily for five days starting 8 hours after the last intravenous Cephradine (Velosef, Bristol-Myers Squibb, Dublin, Ireland). The dogs were reassessed 2, 6 and 12 months after surgery either by direct examination by the same veterinary neurologist or by telephone/email interview. Thereafter the dogs were assessed yearly. The nature of the post operative re-examination depended on owner preference and proximity to the hospital. Nine of 15 dogs had a neurological examination at all time points (unless euthanized). Dog 5 received veterinary neurologist examination at 2, 6, 12 and 23 months only. Dog 10 received veterinary neurologist examination at 2 and 12 months post operatively only. Dogs 3 and 15 received veterinary neurologist examination 2 months post-operatively only. Dog 13 received veterinary neurologist examination 2 months and 2.5 years post-operatively only. Dog 11 had no post-operative examination by a veterinary neurologist with all follow up information being obtained via telephone and email interviews with the primary veterinary surgeon and owner and examination of the primary veterinary surgeon's medical records. All the owners were asked whether they felt that the dog was improved after surgery and questioned on the degree of discomfort displayed by the dog. In the dogs that improved post-operatively the end point was taken as time at which new signs of pain had started to develop and / or additional analgesia was required. However if the post-operative improvement was sustained the end-point was taken as the time after surgery at the last post-operative assessment. In the dogs that were initially unchanged post-operatively an end point was taken as the time that the owner felt that the dog was worse than the preoperative status or if the status was stable as the time after surgery at the last post-operative assessment.

Results

Surgical management

Table 1 illustrates pre-operative information for the 15 CKCS with CM/SM. There were 6 male and 9 female and the age range at the time of first clinical signs of syringomyelia was 0.3-4.5 years (mean 2.2 years). The age at the time of surgery ranged from 0.8-8.1 years (mean 3.3years) and the dogs had clinical signs for 0.1-4.2 years (mean 1.1 years, mediam 0.7y) before surgery. The maximum width of the syrinx ranged from 0.6-0.9cm in 12 dogs (mean 0.7cm). For 3 dogs this information was not available as the MRI images were not in a DICOM ™ format thus preventing precise measurement. These dogs were either assigned wide (2 dogs) or narrow (1 dog) syrinx. Ten of 15 owners made the decision for surgery because they felt that their dog's quality of life was unacceptable; 5 of 15 owners made a decision for surgery in an attempt to avoid deterioration. 1 dog had a pain score of 2, 4 dogs had pain score 3 and 10 had pain score 4. All dogs had a cranial cervical decompression, 8 dogs had a durotomy alone and 7 dogs had durotomy with biocompatible collagen matrix patch (Vet BioSIST, Cook/Global Veterinary Products, SurgiVet, Smiths Medical Pm inc N7 W22025 Johnson Road, Waukesha, WI USA 53186). A decision had been made to change surgical technique because of concern about how the neural tissue bulged through the open dura and arachnoid layers.

Table 1 CKCS with surgical management of Chiari-like malformation / syringomyelia

Dog	Sex	Age onset signs SM (years)	Owner's primary reason for surgery	Pain score	Other neurological signs	Maximum width of syrinx (cm)	Duration clinical signs before surgery (years)	Age at surgery (years)	Surgical technique
1	M	2.6	1	4	Slight thoracic limb weakness	0.7	0.6	3.2	durotomy
2	F	0.3	1	4	Scoliosis, thoracic limb weakness,	wide	1.0	1.3	durotomy
3	F	0.7	2	2	Scoliosis	0.7	0.1	0.8	Durotomy and biocompatible collagen matrix%
4	F	1.5	2	3	Slight thoracic limb weakness	0.8	0.4	1.9	durotomy
5	F	1.7	1	4	Epilepsy, Mild pelvic limb weakness	0.8	1.5	3.2	durotomy
6	F	0.4	1	4	Slight thoracic limb weakness	0.8	1.0	1.4	Partial durotomy *
7	F	3.0#	2	3	none	0.8	1.4	4.4	durotomy
8	M	3.9	1	4	Scoliosis, pelvic limb ataxia, thoracic limb weakness,	0.7	4.2	8.1	Durotomy and biocompatible collagen matrix%
9	F	4.3	1	4	None	0.6	2.8	7.0	Durotomy and biocompatible collagen matrix%
10	M	4.5	2	3	single seizure	0.6	0.6	5.1	Durotomy and biocompatible collagen matrix%
11	F	2.9	1	4	Scoliosis	0.9	0.3	3.3	Durotomy and biocompatible collagen matrix%
12	F	1.3	1	4	Scoliosis	0.8	0.1	1.4	Durotomy and biocompatible collagen matrix%
13	M	3.3	1	4	None	0.7	0.7	4.0	durotomy
14	M	2.1	2	3	Pelvic limb ataxia	narrow	0.8	3.0	Durotomy and biocompatible collagen matrix%
15	M	1.0	1	4	Scoliosis, tetraparesis, ataxia	wide	0.7	1.7	durotomy
MEAN	60%F	2.2		3.6		0.7	1.1	3.3	

F – female, M - male
1. Quality of life unacceptable – pain uncontrolled by medical management.
2. Attempt to prevent deterioration
%-. Biocompatible collagen matrix (Vet BioSIST, Cook/Global Veterinary Products, SurgiVet, Smiths Medical Pm inc N7 W22025 Johnson Road, Waukesha, WI USA 53186).
- clinical signs present from day acquired from breeder at 3 years old
* - problems haemorrhage forced early closure
wide – maximum width of syrinx judged to be more than half diameter spinal cord,
narrow - maximum width of syrinx judged to be less than half diameter spinal cord

Table 2 Post operative follow-up in CKCS with Chiari-like malformation / syringomyelia

Dog	Immediate post operative complications	Apparent dis-comfort Improved after surgery	Pain score 2 months after surgery	Pre-operative neurological deficits improved after surgery	Time of deteriora-tion after surgery (years)	Time after surgery eutha-nasia (years) - reason	Post operative MRI- time (years)	Years since surgery (surviving dogs)	Addi-tional medi-cation #	Age surviving dogs at last follow up (years)
1	none	Y	2%	N	0.2	0.4 - SM	N		none	
2	none	Y	3	Y*	0.3	4.7 - SM	N		S	
3	none	N	2	Y	0.8		N	1.3	None	2.1
4	none	Y	2	N	1.5		Y- 1.0	1.9	F, G	3.8
5	status epilepticus	Y	2	Y	1.8		Y- 1.9	3.8	S	7.0
6	none	Y	2	N	2.0		Y - 2.1	2.4	S	3.8
7	none	N	3	n/a	2.3		Y- 3.8	3.8	G	8.2
8	none	Y	2	Y		0.9 - tonsillar carcinoma			none	
9	Slight ataxia and hypermetria	Y	3	n/a			Y- 0.3	1.0	none	8.0
10		Y	2	seizure 3-4 months post-operatively			N	1.0	F	6.1
11	none	Y	2	No post-operative examination			N	1.2	none	4.4
12	none	Y	2	Y			Y - 1.4	1.6	C,G,A	3.0
13	Slight ataxia and hypermetria	Y	3	n/a			N	2.5	none	6.5
14	none	N	3	Y			N	3.5	none	6.4
15	hypermetric left side, more ataxic	Y	3	Y			N	6.5	none	8.3
MEAN			**2.4**		**1.3**	**2.0**	**1.8**	**2.5**		**5.6**

% - deteriorated within days of this assessment * - improvement of scoliosis only , n/a – not applicable, # - medication dog is currently receiving or was receiving at time of death is detailed F- frusemide, G-gabapentin, C-carprofen, A-acupuncture, S – methylprednisolone or prednisolone , Y – yes, N- no, n/a – not applicable

Table 2 illustrates post operative data for the 15 dogs. The surgery had a low morbidity. One epileptic dog had status epilepticus after surgery and was managed with intravenous diazepam (Diazamuls, Dumex, Barnstaple, UK) at 0.5 mg/kg followed by intramuscular phenobarbitone (Martindale Pharmaceuticles Romford UK) at 6 mg/kg. Three dogs were slightly hypermetric and ataxic the day after surgery. This resolved within 48 hours. All the dogs made a quick recovery after surgery and were able to exercise normally within 4 weeks of the procedure. Six dogs had scoliosis prior to surgery. In 5 dogs this improved; one dog (case 11) had no post operative neurological examination. Four dogs had pelvic limb ataxia with upper motor neuron weakness; this improved in all dogs postoperatively. Six dogs had thoracic limb weakness; this improved in 2 dogs and was persistent in 4. The epileptic dog (case 5) with status epilepticus after surgery never had another seizure post operatively and was successfully weaned of anti-epileptic drugs. Case 10 had a single seizure prior to surgery and another single seizure approximately 3 months after surgery.

None of the dogs had a complete resolution of the signs of pain post operatively. At 2 months post operatively all 10 dogs with a pain score of 4 had improved to pain score 2 (6 dogs) or 3 (4 dogs). Two of the dogs with a pain score of 3 had improved to 2; the remaining 2 dogs were unchanged. The single dog with a pain score of 2 was unchanged post-operatively Overall of the 15 dogs, 12 (80%) had improved comfort after surgery and 7 of that 12 maintained that improvement for 1 – 6.5 years of follow up (mean 2.5 years, median 2.2 years). 1 dog was euthanized because of a tonsillar carcinoma 5 months after surgery; at the time of euthanasia he had maintained an improved status. 4 of the 12 improved dogs had deteriorated 0.2 – 2.0 years after surgery. 2 of these dogs were subsequently euthanized as a consequence of syringomyelia associated pain at 0.4 and 4.6 years after surgery. 2 of 15 dogs had an unchanged pain status postoperatively then subsequently deteriorated 0.8 and 2.3 years after surgery. 1 dog with an unchanged status has remained stable for 3.5 years. In summary all dogs had either an improved (80%) or unchanged status (20%) postoperatively. 7 dogs (47%) subsequently deteriorated 0.2 – 2.3 years after surgery (mean 1.3 years). 2 dogs had been euthanized as a consequence of the syringomyelia and 1 dog because of tonsillar carcinoma; 12 dogs were still alive 1-6.5 years after surgery.

6 of 15 dogs had a post-operative MRI, 0.3-3.8 years (mean 1.8 years) after surgery. Some examples of pre and post operative MRI scans are depicted in figures 5, 6 and 7. It can be appreciated that the shape of the syrinx had remained remarkably unchanged. 7 dogs received additional pain-relief medication after surgery (table 2). Dogs 10 and 12 had been maintained on medication since the time of surgery, the remaining dogs had been started after an increase in discomfort.

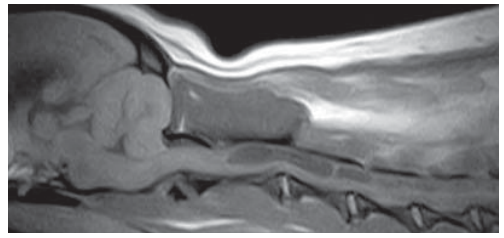

Figure 5a

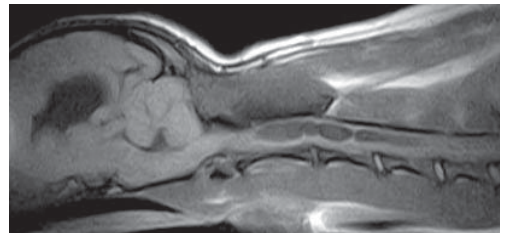

Figure 5b

Figure 5 Midsagittal T1-weighted image of the brain and cervical spinal cord from dog 9 a) pre operatively b) 4 months post operatively. The dog was clinically improved at the time of the MRI scan and described by the owner as no longer screaming. However she still displayed signs of a neuropathic pain syndrome.

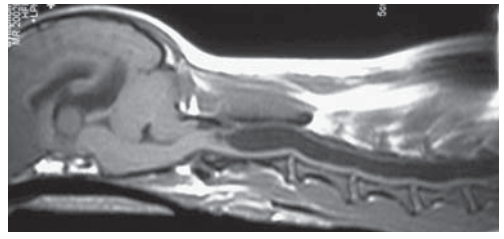

Figure 6a

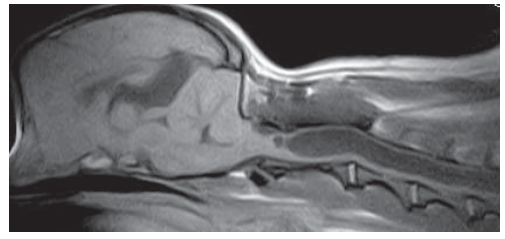

Figure 6b

Figure 6 Midsagittal T1-weighted image of the brain and cervical spinal cord from dog 4 a) pre-operatively b) 1 year post operatively. The dog was clinically improved at the time of the MRI scan however she deteriorated to her pre-operative degree of discomfort 6 months later.

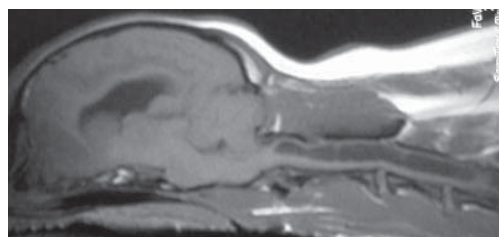

Figure7a

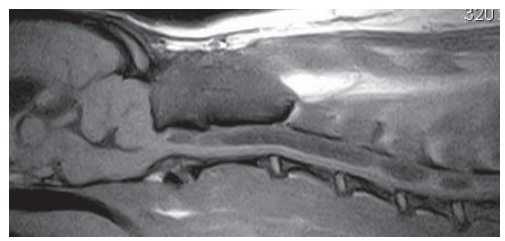

Figure7b

Figure 7 Midsagittal T1-weighted image of the brain and cervical spinal cord from dog 6 a) pre-operatively b) 2 years post operatively. The dog had recently deteriorated to her pre-operative degree of discomfort. The pain was unresponsive to NSAIDS and gabapentin and was somewhat improved on corticosteroids.

Discussion

Successful surgical management of syringomyelia could be defined as a procedure with a low mortality and morbidity where postoperatively the syrinx has resolved and the patient is free of clinical signs and the syrinx does not recur. Like previous reports, this study suggests that cranial cervical decompression is a safe procedure if performed by a surgeon with appropriate neurosurgical training [10, 11, 12]. Intra-operative complications were minimal and confined to meningeal vessel bleeding which was quickly controllable with cautery and haemostatic aids.

The surgery had a low morbidity. All the dogs made a quick recovery after surgery and all were able to exercise normally within 4 weeks after the procedure. 12 of 15 dogs were improved after surgery but all dogs continued to have signs suggesting a neuropathic pain syndrome. All of the dogs with a pain score of 4 improved, suggesting that surgery could be recommended for dogs in severe pain where medical management has been unsuccessful. However although 80% of the dogs showed a post operative improvement this surgical procedure does not appear to result in resolution of the syrinx. Similar findings were described by Vermeersch et al [11]. The reason for the failure for this surgical technique to adequately deal with syringomyelia is not clear. Da Costa, et al described successful management of acquired cerebellar herniation and syringomyelia secondary to brain stem tumour [13]. After treatment with radiation therapy, shrinkage of the tumour and presumably re-establishment of CSF flow through the foramen magnum the syringomyelia resolved indicating that reversal is possible in the dog. The failure of surgery to adequately treat the syrinx suggests that this surgery in the dog does not sufficiently improve CSF flow though the foramen magnum and it may be that in the dog, removing most of the supraoccipital bone and part of the dorsal laminae of C1 with durotomy is just not sufficient. The same situation does not exist for management of Chiari malformation in humans in which most reports suggest that cranial cervical decompression, whatever the technique, results in syrinx resolution in over 90% of patients [14, 15]. However due to anatomical differences it is possible to remove comparatively more bone laterally from both the occipital bone and atlas in the human. One further difference between the human and canine condition is that in the human, the cerebellar tonsils rather than the vermis is herniated through the foramen magnum[16]. The tonsils may be resected without compromise to the patient and consequently many surgeons partly or completely remove them thus creating more space[17, 18]. However, resection of the vermis in the dog could be expected to result in post-operative ataxia, dysmetria, and intention tremors[19]. It may be possible to perform a partial resection of the vermis without serious postoperative complication and this it may be worth considering as an adaptation of the surgery (Todd Axlund, personal communication).

Another possible explanation is that once formed a large syrinx may self perpetuate either because normal CSF flow cannot be re-established though the narrowed subarachnoid space or possibly because the CKCS spinal cord with chronic syringomyelia is not compliant enough to allow syrinx collapse and resolution. An alternative explanation is that pathogenesis of the syringomyelia in the CKCS is due to more than just overcrowding at the foramen magnum. Chiari malformation is extremely common in the CKCS [5,6]. Cerda-Gonzalez et al found that 51 of 59 dogs had cerebellar crowding at the foramen magnum[20]. However, not all dogs with a Chiari malformation develop syringomyelia and there is no significant difference in the caudal fossa volume between CKCS with and without syringomyelia.[20, 21] This suggests that there may be other anatomical factors involved; so far however, investigation has failed to indicate what these may be. In a study looking at vertebral canal diameter in the C1-C3 area, Carruthers et al [21] found that dogs with syringomyelia tended to have a larger vertebral canal diameter at the C2/C3 junction and at C3 and that dogs with syringomyelia and clinical signs of pain tended to have a narrower vertebral canal at C1/C2 although further study is required to establish if this truly is an important contributory feature in the pathogenesis.

It is possible that the surgery could be modified and improved. Dewey et al recently described a technique of cranial cervical decompression combined with a titanium polymethyl methacrylate plate over the bony defect. [22] It is not yet known whether this technique will be more successful in the long term. An alternative method of managing syringomyelia is direct shunting of the cavity. In humans this is not a preferred technique for management of CM/SM as long term outcome is poor due to shunt obstruction and/or spinal cord tethering.[23] However the short term results are reasonable with syrinx collapse occurring in a majority of human cases[24]. There has been a single report of syringo-subarachnoid shunting in a dog using an equine ocular lavage tube (Cook/Global Veterinary Products, SurgiVet, Smiths Medical Pm inc N7 W22025 Johnson Road, Waukesha, WI USA 53186). [25] However post-operative MRI revealed that the syringomyelia was still prominent although there was clinical improvement.

7 of 15 dogs (47%) deteriorated 0.2-2.3 years after surgery (mean 1.3 years). The follow up period of the remaining dogs ranged from 1 to 6.5 years (mean 2.5 years) so more cases could be expected to deteriorate given a longer follow up period. Similar results are reported by Dewey et al. [10] In a series of 16 dogs, 13 (81%) of affected dogs had improvement of clinical signs after surgery, but 25% had a recurrence within the follow-up period of 6-36 months.[10] It was not ascertained whether any of the improved dogs actually had resolution of the syrinx prior to deterioration or were in fact similar to the dogs in the current study i.e. clinical improvement without radiographic improvement. The deterioration was attributed to scar tissue adhering to exposed neural tissue thus preventing adequate CSF flow. [10] In the current study 1 of the 2 dogs that was euthanized after an early recurrence of signs was necropsied (Dog 1). Gross inspection revealed many layers of scar tissue adhered to the dorsal medulla, cranial spinal cord and cerebellum. This author suggests that the post operative improvement after cranial cervical decompression in canine CM/SM is attributable to improved CSF flow at the foramen magnum and the deterioration is due to subsequent obstruction.

It is possible that although cranial cervical decompression in the dog does not reverse the syrinx it may delay the progression of the disease. However due to the small numbers in this series it is not

possible any draw any conclusions as to whether the surgery did in fact achieve this objective. The post-operative MRI scans demonstrated little or no improvement in the syringomyelia but they also showed no progression. However it is naive to assume that because the syrinx had not increased in size and / or the dogs euthanized as a consequence of pain that the surgery was a partial success as the natural progression of CM/SM without surgical management has not been established. This also raises some ethical issues as syringomyelia results in a neuropathic pain syndrome [8] and according to human suffers, is an extremely painful condition. [7] Merely surviving does not give an indication of quality of life or whether the discomfort these animals experience is appropriately addressed.

The majority of the dogs in this study had high pain scores (mean score 3.6) and wide syrinxes. It is not known if this surgery may be more successful if performed on dogs with less severe disease. Due to small study size it was also not possible to draw conclusions as to whether performing surgery earlier in the course of the disease was advantageous however there was no correlation between favourable outcome and short clinical history of signs of syringomyelia.

As this is a small retrospective clinical study with many methodological problems, conclusions, if any, must be drawn with caution. One of the most important problems with this study is that pain is a subjective parameter and may have been inappropriately assessed. In particular interpretation of clinical history such as “scratching less intensely and less often” was very subjective and the pain score used in the study, although helpful, was not detailed enough. An improved (but simple) way of prospectively scoring neuropathic pain in dogs needs to be established (appendix 1). In an attempt to avoid bias the end point of this study was taken to be when the owners felt that the dog was starting to show new signs of pain, for example starting to scream more frequently. However this parameter is also subjective especially as owners may become more tolerant of clinical signs as they get used to them. Another limitation of this study is that not every dog had a postoperative MRI and with 3 dogs the last post operative assessment(s) was done distantly by telephone and/or email and examination of medical record.

Conclusion

Cranial cervical decompression surgery for CM/SM has a low mortality and morbidity and in cases with a high pain score the procedure can improve quality of life. The procedure does not appear to result in syrinx collapse and resolution possibly because other factors are involved in the development and/or persistence of syringomyelia. In addition the clinical improvement may not be sustained and a proportion can be expected to deteriorate from as soon as 2 months after surgery. The procedure may or may not delay progression of the disease. It is suggested that the postoperative improvement is attributable to improved CSF flow at the foramen magnum and the deterioration is due subsequent obstruction. It is also suggested that to properly evaluate the success of any surgical technique for CM/SM that there be a follow up period of at least 2 years and that post operative MRI scans be performed as clinical improvement does not necessarily imply radiographic improvement. Further study is needed to achieve better understanding of the pathogenesis and predisposing factors of CM/SM in the dog. As this condition is painful but does not necessarily result in euthanasia better medical treatment guidelines should be established.

Acknowledgements

The author is grateful to Penny Knowler who prepared Figures 2 and 4 and to the Goddard Veterinary group who part financed most of the post-operative MRI scans. I am also grateful to Professors Nick Jeffery and Jan Rothuizen who critically reviewed the manuscript.

References

1. Rusbridge C, Greitz D, Iskandar BJ. Syringomyelia: Current concepts in pathogenesis, diagnosis and treatment. J Vet Intern Med 2006; 20: 469-79.
2. Rusbridge C, MacSweeny JE, Davies JV et al. Syringomyelia in Cavalier King Charles Spaniels. J Am Anim Hosp Assoc 2000; 36: 34-41.
3. Rusbridge C, Knowler SP. Hereditary aspects of occiptial bone hypoplasia and syringomyelia (Chiari type I malformation) in cavalier King Charles spaniels, Vet Rec 2003;153:107-12.
4. Rusbridge C, Knowler SP. Inheritance of Occipital Bone Hypoplasia (Chiari type I malformation) in Cavalier King Charles spaniels. J Vet Intern Med 2004; 18: 673-8.
5. Greitz D. Unravelling the riddle of syringomyelia. Neurosurg Rev 2006; 29:251-63.
6. Levine DN. The pathogenesis of syringomyelia associated with lesions at the foramen magnum: a critical review of existing theories and proposal of a new hypothesis. J Neurol Sci 2004; 220: 3-21.
7. Todor DR, Harrison TM, Milhorat TH.. Pain and syringomyelia: A review. Neurosurg focus 2000; 8: 1-6.
8. Rusbridge C, Caruthers H, Dubé M-P, et al Syringomyelia in cavalier King Charles spaniels: the relationship between syrinx dimensions and pain J Small Anim Pract 2006; In Press
9. Woolf CJ. Dissecting out mechanisms responsible for peripheral neuropathic pain: implications for diagnosis and therapy. Life Sci 2004; 74: 2605-10.
10. Dewey CW, Berg JM, Barone G et al. Foramen magnum decompression for treatment of caudal occipital malformation syndrome in dogs. J Am Vet Med Assoc 2005; 227: 1270-5.
11. Vermeersch K, Van Ham L, Caemaert J et al. Suboccipital Craniectomy, Dorsal Laminectomy of C1, Durotomy and Dural Graft Placement as a Treatment for Syringohydromyelia with Cerebellar Tonsil Herniation in Cavalier King Charles Spaniels Vet Surg 2004; 33: 355-60
12. Churcher RK, Child G. Chiari 1/syringomyelia complex in a King Charles Spaniel. Aust Vet J. 2000; 78: 92-5.

13. da Costa RC, Parent JM, Poma R, Duque MC. Cervical syringohydromyelia secondary to a brainstem tumor in a dog. J Am Vet Med Assoc. 2004; 225:1061-4.
14. Sacco D, Scott RM. Reoperation for Chiari malformations. Pediatr Neurosurg. 2003; 39:171-8
15. Tubbs RS, McGirt MJ, Oakes WJ. Surgical experience in 130 pediatric patients with Chiari I malformations. J Neurosurg. 2003; 99:291-6.
16. Cai C, Oakes WJ.Hindbrain herniation syndromes: the Chiari malformations (I and II). Semin Pediatr Neurol. 1997;4:179-91.
17. Guyotat J, Bret P, Jouanneau E, et al. Syringomyelia associated with type I Chiari malformation. A 21-year retrospective study on 75 cases treated by foramen magnum decompression with a special emphasis on the value of tonsils resection. Acta Neurochir (Wien). 1998; 140:745-54
18. Fischer EG. Posterior fossa decompression for Chiari I deformity, including resection of the cerebellar tonsils. Childs Nerv Syst. 1995;11:625-9.
19. Kornegay JN. Cerebellar vermian hypoplasia in dogs. Vet Pathol. 1986; 23:374-9.
20. Cerda-Gonzalez S, Olby NJ, Pease TP, et al. Morphology of the Caudal Fossa in Cavalier King Charles Spaniels. J Vet Intern Med 2006; 20: 736
21. Caruthers H, Rusbridge C, Dubé, M-P, et al Association between cervical and intracranial dimensions and syringomyelia in the cavalier King Charles spaniel J Small Anim Pract 2006; Submitted
22. Dewey CW, Bailey KS, Marino DJ, et al. Foramen magnum decompression with cranioplasty for treatment of caudal occipital malformation syndrome is dogs. J Vet Intern Med 2006; 20: 783.
23. Medow J, Sansone J, Iskandar BJ, Syringomyelia and Hydromyelia In: Albright AL, Pollack AF, Adelson P.D (eds) Principles and Practice of Pediatric Neurosurgery 2nd Edition, Thieme Medical Publishers 2006; Awaiting publication
24. Iwasaki Y, Hida K, Koyanagi I, et al. Reevaluation of syringosubarachnoid shunt for syringomyelia with Chiari malformation. Neurosurgery. 2000; 46: 407-12.
25. Skerrit GC, Hughes D. A syndrome of syringomyelia in the cavalier King Charles spaniel, and its treatment by syringo-subarachnoid shunting. In Proceedings from the 12th Annual Symposium of the European Society of Veterinary Neurology, Vienna, 1998; 23

Chapter 6.2

Pilot Study: Chiari-like malformation with syringomyelia in the cavalier King Charles spaniel; long term follow up after conservative management

C Rusbridge

Stone Lion Veterinary Centre, 41 High Street, Wimbledon Village, London, SW19 5AU

Introduction

The progression of syringomyelia without surgical management has not been documented and without this background information it is more difficult to assess success or otherwise of surgery. A pilot study looking at the natural history of syringomyelia in a group of cavalier King Charles spaniels (CKCS) displaying signs of neuropathic pain syndrome was therefore performed.

Materials and methods

The medical records of all cavalier King Charles spaniels (CKCS) presenting to the Stone Lion Veterinary Centre neurology service over a 5 year period and with at least 2 years follow up (July 1999 to July 2004) were examined for a diagnosis of syringomyelia or suspected syringomyelia. The records were further searched for a clinical history of phantom scratching.[1] This clinical sign was chosen because it is specific for syringomyelia and is an indication of a wide syrinx with a neuropathic pain syndrome, thus ensuring that all the dogs in the group had disease of a similar severity[1] A further advantage was that this allowed a comparison with a similar group of CKCS that were managed surgically.[2] The owners and veterinary surgeons for this sub group of dogs were contacted by telephone and / or email and sent a questionnaire (Figure 1) detailing the dog's clinical signs and treatment. The veterinary surgeons also supplied a copy of their medical records for each dog. The end point was taken as the age at the point of death or the age at the conclusion of the study in July 2006.

Figure 1 Questionnaire sent to owners and veterinary surgeons (1 copy each) of dogs with clinical signs of syringomyelia-associated neuropathic pain ▶

The cohort included 6 dogs without MRI (magnetic resonance imaging) confirmation so as not to exclude dogs from a different socioeconomic group (i.e. owners that could not afford a MRI scan) and because the perception of suffering and decision making may be different between owners allowing and refusing investigation.

Results

Twenty eight dogs met the criteria for inclusion in the study, of which 11 were managed surgically and 17 conservatively. 3 of those conservatively managed dogs were lost to follow up because the owners had moved and the details of the 14 remaining dogs are detailed in Table 1. The age at which first signs of syringomyelia were seen was 0.4 – 6.8 years (mean 2.2 years) and the age of diagnosis was 1.3 – 9.1 years (mean 4.4 years). 8 of 14 dogs had the suspected diagnosis confirmed by MRI or post mortem.

Treatment included nonsteroidal anti-inflammatory drugs (NSAIDs), gabapentin (Neurontin Parke-Davis, Eastleigh, UK) and corticosteroids but some dogs received no drugs at all. There was great variation in the treatment regime between individuals at any given time but typically dogs were initially started on NSAIDs and then switched to gabapentin and / or steroids as their condition progressed. Five dogs were euthanized because of syringomyelia associated pain at 3.0-10.4 years old (mean 6.3 years), one dog was euthanized because of heart failure and the remaining dogs are still alive aged 4.1 – 10.3 years old (mean 7.1 years). By comparison in the group of 15 dogs managed surgically,[2] there was an post operative improvement in 80% with the remainder unchanged. Seven dogs (47%) subsequently deteriorated 0.2 – 2.3 years after surgery (mean 1.3 years) and 2 dogs were eventually euthanized as a consequence. Twelve dogs were still alive, 1-6.5 years after surgery (mean 2.5 years).

CKCS SYRINGOMYELIA STUDY GROUP

Aiming to prevent pain in our dogs

Pedigree Name / Registration Number _____

Dog's Date of birth: _____ Colour B B/T R T Gender M, MN F FN Date questionnaire completion_____

CLINICAL SIGNS

At what age did your dog first show signs of syringomyelia? _____

What were the initial clinical signs?

Shoulder scratching ☐; Scratching elsewhere☐ (specify)_____ ; Neck pain ☐ ; Pain elsewhere ☐ Specify _____ ; Signs of pain when scratching ☐, excited ☐, touched ☐, change of head position ☐, jumping ☐, no apparent reason ☐; Scoliosis (twisted spine esp. neck) ☐ Wobbly hind limb gait ☐ Weak forelimbs ☐

When was your dog diagnosed with syringomyelia?_____How old was your dog?_____

How was the diagnosis made? MRI ☐ Post Mortem ☐ Suspected on basis clinical signs only ☐

If possible, please attach a copy of the MRI or PM report / findings.

If applicable, what were the clinical signs before surgery?

Shoulder scratching ☐; Scratching elsewhere☐ (specify)_____ ; Neck pain ☐ ; Pain elsewhere ☐ Specify _____ ; Signs of pain when scratching ☐, excited ☐, touched ☐, change of head position ☐, jumping ☐, no apparent reason ☐; Scoliosis (twisted spine esp. neck) ☐ Wobbly hind limb gait ☐ Weak forelimbs ☐

Is your dog alive? Yes ☐ No ☐ **If dead, at what age did they die?**_____

If dead, what was the cause of death / reason for euthanasia?_____

_____

What are dog's clinical signs now (if dead indicate clinical signs at time of death)

Shoulder scratching ☐; Scratching elsewhere☐ (specify)_____ ; Neck pain ☐ ; Pain elsewhere ☐ Specify _____ ; Screaming when scratching ☐, excited ☐, touched ☐, change of head position ☐, jumping ☐, no apparent reason ☐; Scoliosis (twisted spine esp. neck) ☐ Wobbly hind limb gait ☐ Weak forelimbs ☐

TREATMENT☐ Please Complete Page 2 NO TREATMENT☐

Please return questionnaire to CKCS syringomyelia project coordinator Clare Rusbridge, Stone Lion Veterinary Centre, 41 High Street, Wimbledon, UK, SW19 5AU neuro.vet@btinternet.com Confidential fax 00 44 (0)20 87860525

CKCS SYRINGOMYELIA STUDY GROUP

Aiming to prevent pain in our dogs

Treatment	When / how old was dog	How long did your dog have this treatment	Was it effective? (PLEASE GRADE 0-5 WHERE 0 IS COMPLETELY INEFFECTIVE AND 5 IS COMPLETELY EFFECTIVE)	How long was it effective for	Is your dog still receiving this drug
NSAIDS e.g. Rimadyl, Metacam					
Steroids e.g. prednisolone					
Gabapentin (Neurontin)					
Opioid drugs e.g. pethidine or morphine					
Acetazolamide (Diamox)					
Shunt surgery (syrinx to subarachnoid shunting)		N/A			N/A
Decompression surgery (occipital craniectomy +/- durotomy, C1 laminectomy)		N/A			N/A
Repeat shunt surgery		N/A			N/A
Repeat decompression surgery		N/A			N/A
Acupuncture					
Homeopathy					
Other (please specify)					

If dog had surgery please indicate Surgeon's name:_____Phone:_____

Address:_____

Please return questionnaire to CKCS syringomyelia project coordinator Clare Rusbridge, Stone Lion Veterinary Centre, 41 High Street, Wimbledon, UK, SW19 5AU neuro.vet@btinternet.com Confidential fax 00 44 (0)20 87860525

Table 1 14 CKCS with signs of syringomyelia associated neuropathic pain

DOG	Sex	Age onset signs SM (years)	MRI / PM confirmed	Age diagnosis SM (years)	Age at death (years)	Reason for euthanasia	Years with clinical signs SM	Age surviving dogs at last follow up (years)
1	M	2.3	Y	2.7	3.0	sm	0.7	
2	F	1.0	N	3.0	5.2	sm	4.2	
3	F	1.7	Y	3.0	6.3	sm	4.6	
4	F	6.8	N	7.9	9.2	sm	2.4	
5	M	0.5	Y	4.0	10.0	heart failure	9.5	
6	M	1.5	Y	9.1	10.4	sm	8.0	
7	M	1.0	N	1.3			3.1	4.1
8	F	1.0	Y	2.2			3.2	4.2
9	M	1.4	N	1.9			3.1	4.5
10	F	0.4	Y	2.4			5.3	5.7
11	M	6.0	N	7.0			2.6	8.6
12	M	1.5	Y	5.2			5.7	9.5
13	M	1.0	Y	4.0			9.0	10.0
14	M	4.0	N	8.0			6.3	10.3
MEAN	**36%F**	**2.2**	**57% Y**	**4.4**	**7.3 (6.3 excluding case 5)**		**4.8**	**7.1**

M - male, F – female, Y- yes, N- no, SM- syringomyelia

Discussion

The age of onset and distribution of sex was similar to other studies [2] CM/SM was a cause of euthanasia in 36% of the cases. However 43% of the cohort had survived to be greater than 9 years of age. A recent study suggested that the mean age of death for CKCS is 10.7 years (standard deviation 2.9 years), [3] indicating that syringomyelia does not necessarily result in premature death and many dogs can achieve a normal lifespan for the breed. However, survival does not imply that the dogs enjoyed a good quality of life and the medical records of many of the dogs suggested that the pain was not adequately controlled all of the time. This observation is important when attempting to assess the success (or otherwise) of surgical management and also emphasises that surgery should not be considered a success because the dog has not been euthanized.

This study had serious methodological problems meaning that accurate conclusions cannot be drawn however it suggests that dogs with neuropathic pain from syringomyelia may survive for many years and are likely to benefit from better medical treatment guidelines. It also highlights the need for a prospective clinical study assessing which analgesics are the most appropriate for treating this disorder.

Acknowledgements

The author is grateful to Lisa Fox VN who spent considerable time coordinating much of the telephone and questionnaire follow up for the conservatively managed cases. Thanks are also due to Drs Pete Smith and Paul Mandigers who critically appraised this manuscript.

References

1. Rusbridge C, Caruthers H, Dubé M-P, et al Syringomyelia in cavalier King Charles spaniels: the relationship between syrinx dimensions and pain J Small Anim Pract 2006; In Press
2. Rusbridge, C Long term follow up after surgical management of canine Chiari malformation with syringomyelia, Veterinary Surgery, Submitted 2006
3. Glickman L, Raghavan M, Glickman N. American Cavalier King Charles Spaniel Club, Inc. Health Survey 2004-2005 A Collaborative Effort between ACKCSC Charitable Trust, ACKCSC Health Committee, Board of Directors and Members, ACKCSC, Inc. and Purdue University School of Veterinary Medicine Section of Clinical Epidemiology. Purdue University School of Veterinary Medicine 725 Harrison Street West Lafayette, IN 47907-2027. 2005; 17 http://ackcsc.org/health/ckcshealthsurveyfinalreport.pdf Assessed 8th July 2006