Canine Chiari & Syringomyelia

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• What is syringomyelia?
• Clinical signs of syringomyelia
• Diagnosis of Chiari and syringomyelia
• Treatment of syringomyelia
  – Surgery
  – Medial - covered in next lecture
• Genetics of syringomyelia
• Prevention of syringomyelia
Chiari malformation and syringomyelia

www.inkymousestudios.com
Syringomyelia
obstruction of cerebrospinal fluid movement

Choroid Plexus - produces CSF

CSF flow

Arachnoid granulations – absorb CSF

Foramen magnum
The skull is a box containing the brain, blood vessels and cerebrospinal fluid.
Blood arrives from heart

Blood vessels

Brain

Skull

CSF

C

S

F
Blood returns to heart

Blood vessels

Brain

Skull

CSF
Brain

Blood vessels

Chiari Skull
Blood arrives from heart

Blood vessels

Raised pressure

Brain

Chiari Skull

CSF

Jet of fluid
Pathogenesis of syringomyelia

CSF PULSE PRESSURE

ARTERIAL PULSE PRESSURE

Syringomyelia: A review of the biomechanics
N.S.J. Elliott a, C.D. Bertram b,*, B.A. Martin c, A.R. Brodbelt d
Chiari-like malformation and syringomyelia
"Before you go any further, let me reiterate that I, for one, see nothing wrong with killing the messenger."
What breeds are affected by Chiari and/or syringomyelia?

1. Cavalier King Charles Spaniels
2. King Charles spaniels
3. Griffon Bruxellois
4. Weimaraners
5. Brachycephalic toy dogs
Breeds CM/SM

- Any toy breed dog (and their crosses)
- Especially
  - Cavalier King Charles spaniels
  - King Charles spaniels
  - Griffon Bruxellois
  - Affenpinscher
  - Maltese
  - Yorkshire terrier
  - Pomeranian
  - Chihuahua
  - Papillion
  - Boston terrier
  - Also one SBT
What is the most common clinical sign of Chiari and/or syringomyelia?

1. Progressive paralysis
2. Seizures
3. Balance problems
4. Pain
5. All of the above
Clinical signs – CM/SM

Before medication

After medication

Pain face
Unusual sleeping posture / interrupted sleep
Postural pain

- Pain on being lifted
- Unwilling to jump
- Unwilling to do stairs
Poor exercise tolerance
CM associated pain

Cartoons - Thanks to Tania Ledger (www.cavaliermatters.org)
CM associated pain

Chiari-like malformation associated pain
Dogs in pain are more likely to have a wide syrinx with dorsal grey column damage.

- **Not Painful**
- **Painful**
Shoulder scratching

Especially

• Walking
• Excited
• Touched
• “clothing”
  – neck collar
  – snoods
• Breeders call “air guitar” or “phantom”

Picture Jemima Harrison
thanks Jemima Harrison Passionate Productions
Pedigree Dogs Exposed
Scoliosis / Torticollis

Dorsal grey column damage
more likely with wide syrinxes

Imbalance afferent proprioceptive information from cervical neuromuscular spindles
i.e. Imbalance information telling brain which way is “up”
Does pain affect your outlook on life?

• **Fear of strangers**
• **Non-social fear**
  – unfamiliar situations
  – sudden loud noises
• **Attachment behaviour**
  – ‘clingy’
  – separation-related
• **Excitability**
  – attention-seeking
  – more excitable

Owners’ perception - poorer quality of life

Pain associated aggression
Chiari malformation

Angel 4y FS CKCS – MRI
Syringomyelia

Neurological deficits in more severe cases

2 year old female CKCS
Diagnosis

MRI is only method confirming CM/SM

Syringomyelia is a late onset disease

Screening from 12m (ideally repeated at 5y & in between)
What is the prognosis for Chiari and syringomyelia?

1. Hopeless - euthanize if diagnose
2. Very poor
3. Guarded
4. Good
5. Depends....not willing to commit
Prognosis - Medical management

- Guarded if...
  - wide syrinx
  - first clinical signs before 4 years of age.
- Signs progress in ¾ CKCS with CM+-SM despite medical treatment (study of 48 dogs)
  - Many still enjoy reasonable quality of life
  - 15% euthanatised CMSM
  - Live with CMSM and die from MVD

Plessas IN, et al. Long-term outcome of Cavalier King Charles spaniel dogs with clinical signs associated with Chiari-like malformation and syringomyelia. The Veterinary record. 2012. doi:10.1136/vr.10044
Treatment of syringomyelia

Picture Henny Van De Berg
Cranial cervical decompression

![Image of a dog undergoing cranial cervical decompression procedure.](image-url)
Prognosis after surgery

Surgery usually successful at reducing the pain but...

- Syringomyelia is still present!
- May still have discomfort /scratching
- ~ 50% deteriorate by 2.5y post-op
Cranial cervical decompression

Bone removed from skull & atlas

Meninges is cut and tacked back

Some advocate leaving open, some graft dura, others cover bony defect (cranioplasty)
After FMD - pre and post op MRI

Pre-op

4 months post-op
If associated hydrocephalous -shunting ventricles

B4 intraventricular shunt

15 months post surgery
Any questions?
Genetics

Picture Henny Van De Berg
Inheritance of SM

• Moderately high hereditability
  – SM = 0.37 (maximum is 0.64)
• What does that mean?
  – Complex inheritance involving >1gene.
    • ? genes at two or more loci interact to give disease
  – Expression of disease may be influenced by other as yet unknown factors
  – There is a good chance of “breeding it out”

Genetics of CM

- 2 DNA regions strongly associated with CM
- One excellent candidate gene Sall-1
  - involved in development of skull
  - Mutation Sall-1 in humans
    - Townes-Brocks syndrome
      - Can have CM
    - Branchio-oto-renal syndrome

Quantitative Trait Loci (QTL) Study Identifies Novel Genomic Regions Associated to Chiari-Like Malformation in Griffon Bruxellois Dogs

Philippe Lemay, Susan P. Knowler, Samir Bouasker, Yohann Nédélec, Simon Platt, Courtenay Freeman, Georgina Child, Luis B. Barreiro, Guy A. Rouleau, Clare Rusbridge, Zoha Kibar
Prevalence (CKCS)

KC / BVA CMSM MRI Scheme

Picture Henny Van De Berg
How are the images graded?

- Severity of CM (graded 0, 1, 2)
- Severity of SM (graded 0, 1, 2)
  - If appropriate, size of syrinx – transverse width
- Age at time of scanning \((a(\geq 5y), b(3-5y), c(1-3y))\)
Mate select program (EBV)

Rosie “selects” Winston (photo Carol Fowler)

Official scheme
- Allows clear dogs to be identified
- Is a (minimum) standard / quality control
  - 2 or more scrutineers
  - Many breeders have relied on results of scans that are not good enough
- It takes experience to interpret CMSM
- Feeds into an EBV
“I will never breed to an ugly CKCS even if it is SM clear”

King Charles spaniel with severe syringomyelia that breeder really would like to breed

SM clear “ugly” CKCS that “wouldn’t be used if it was the last dog on the planet”

Which dog is ugly?
Take home message

• Syringomyelia is a disorder of CSF
• CMSM is common in some breeds
• The most important clinical sign is pain
  – Behaviour
  – History
• CMSM a complex inheritance
• BVA KC (and other) breeding schemes
Thank you for listening!

Any questions?
www.veterinary-neurologist.co.uk