The run-through...

- Anatomy and physiology
  - Ventricles and CSF

- Pathophysiology
  - Obstructions and theories

- Diagnosis

- Treatment
  - Medical
  - Surgical
  - Complications
  - Outcomes
Anatomy and Physiology

Brain and CSF
Brain – Ventricular System

A

Lateral ventricle
Mesencephalic aqueduct
Lateral recess, IV ventricle
Central canal
Fourth ventricle
Suprapineal recess, III ventricle

Interventricular foramen

Cavity of olfactory bulb

B

Rostral horn, lateral ventricle
Mesencephalic aqueduct
Caudal horn, lat. vent.
Fourth ventricle
Central canal

Cavity of olfactory bulb

Third ventricle
Optic recess
Infundibular recess

Ventral horn (temporal), lat. vent.
Brain – Ventricular System

- Choroid plexus
  - Band of clustered villi
  - Project into the ventricle
  - Each villus
    - microvascular proliferation
    - cuboidal ependymal lining
  - Production of CSF
CSF – composition/production

- **Composition**
  - Alkaline
  - Positive charge

- **Production**
  - 0.03-0.5ml/min
  - “Replaced” 3-4 times daily
  - 2 methods:
    - Coroid plexus (carbonic anhydrase dependent but independent of ventricular pressure)
    - ECF from brain and spinal cord
CSF - Flow

Lateral ventricles

Interventricular foramina

3rd Ventricle

Mesencephalic aqueduct

4th Ventricle  Central canal

Lateral recesses and apertures

Subarachnoid space
CSF - Flow
CSF - Absorption

- **Route 1:**
  - **Arachnoid Villi**
    - Into venous sinus
  - Flat fibroblasts & Endothelial cells
  - Intercellular spaces open on expansion
    - 7-10 cmH\(_2\)O
  - Also at intervertebral foraminal veins
CSF – Absorption

Route 2:
- Meningeal sheaths
  - Meningeal sheaths around nerve roots
  - At the distal termination of meninges CSF “leaks”
  - Escapes into peripheral nerve lymphatics
Pathophysiology

Congenital or acquired, ventricular or extraventricular, communicating or not?
Pathophysiology - Nomenclature

• Hydrocephalus ex vacuo
  • Infarcts, necrosis may leave a vacant space which fills with CSF rather than active distension
  • Old terminology and now not classified as hydrocephalus at all

• Communicating or not?
  • Subarachnoid space and ventricular space
  • Walter Dandy – early 1900s

• Congenital or Acquired?

• Intraventricular or Extraventricular
  • The current classification system (Rekate 2009)
Pathophysiology

- **Rate of formation versus rate of absorption**
  - Formation constant
  - Pressure gradient sets off development of hydrocephalus
    - $<0.5 \text{ mmHg}$ (Levine 1999, 2008)
    - Alternate pathways can develop
      - CSF flow across ependyma to capillaries
      - CSF flow across cribiform plate to nasal lymphatics

- **Hydrodynamic theory** (Greitz 2004)
  - Proposes hydrocephalus a result of reduced brain compliance and the effect this has on brain capillaries
    - Normally brain capillaries don’t pulse but if they do, the increased pressure during pulse phase causes a transmantle pressure gradient and fluid flow
    - May explain “normal pressure hydrocephalus”
Pathophysiology - ICP

• 3 factors determine whether hydrocephalus causes ICP rise:
  1. Size of pressure gradient
     ▪ Severity of obstruction
     ▪ Alternate pathway development
  2. Efficiency of pressure transfer
     ▪ Brain elasticity -> incompressible early in hydrocephalus, brain “softens” later in disease
  3. Ventricular size
     ▪ Smaller ventricles = more cortical matter
     ▪ Greater cortical mass is more resistant to ventricular pressure
Intraventricular obstruction

- Unilateral ventricle
  - mass or inflammatory lesion

- 3rd ventricle mass
  - both lateral ventricles dilate

- Mesencephalic duct
  - inflammatory or developmental (toys)
  - can be secondary to temporal horn enlargement (worsening signs)

- Lateral apertures
  - developmental (Chiari?), Tumours, inflammation (FIP)
Subarachnoid obstructions

- Intraventricular haemorrhage
  - Human premature babies

- Occlusion and agenesis of arachnoid villi
  - Meningitis
  - Developmental defect
  - Ventricles and subarachnoid space distend
Pathophysiology – brain damage

- Vascular damage
- Compression
  - Ependyma cells flatten and lose cilia
  - Ependymal integrity loss
    - Periventricular white matter oedema
    - White matter direct compression
  - Structural loss
    - Septum pellucidum, cortical loss if severe
    - Cortical damage critical to prognosis following shunting
Pathophysiology – brain damage

Thinning of white matter

Septal defects
Diagnosis

Clinical features and diagnostic imaging
Clinical Features & Exam Findings

- Congenital
  - Toys breeds
    - Mesencephalic aqueduct stenosis common
    - Subarachnoid / Arachnoid villi
    - Transient stenosis?
    - Genetic factors?
    - Infection?
    - Other anomalies – Chiari, cerebellar hypoplasia, Dandy-Walker syndrome, etc
Clinical Features & Exam Findings

- Signs (Congenital)
  - See image!
  - Head shape & strabismus
  - Runts
  - Neuro changes
    - Abnormal cognition
    - Altered consciousness & seizures
    - Ataxia, circling, blindness (very commonly), vestibular changes
  - Variable signs and degrees of changes
  - May be static, worsen or sometimes improve with time
  - Can present as young adults
    - 205/564 (36%) of dogs classified with congenital hydrocephalus were >2 years old (Selby AJVR 1979)
Clinical Features & Exam Findings

- Signs (Acquired)
  - Any age
  - Range of neuro signs similar to congenital form
  - Signs may reflect underlying cause
    - Tumours, meningitis
  - Note: cranial sutures have closed so skull can’t deform
Ventricular size

- Measured at level of interthalamic adhesion
- **Lateral** ventricles deemed large if:
  - (Hudson 2005)
    - Ventricular height >0.35cm
    - or,
    - Ventricular height to cerebral width ratio >0.19
  - (Spaulding 1990)
    - Ventricular height to dorso-ventral cerebral height >0.14
Diagnosis – Imaging

- But....
  - There is a poor correlation between ventricular size and clinical signs
  - Asymmetric enlargement of lateral ventricles is common in normal brains

- Therefore....
  - Diagnosis based on clinical signs and imaging not imaging alone
Ultrasound

- High frequency probe (7-12MHz)
- Only with open/persistent bregmatic fontanelle
- Good for ventricular size and ongoing monitoring but resolution inferior to CT or MRI
CT and MRI

- Closed fontanelles not a problem
- Ventricular size
- Assessment of cortical atrophy
- Identify focal lesions
  - Assess all ventricles and subarachnoid space together
- Post-shunt placement
- Ventricular oedema
- Assess cerebellomedullary cistern before CSF tap
Diagnosis - Imaging
Diagnosis

- **CSF analysis**
  - May help identify inflammation
  - Check cistern is full of brain first....
  - May be safer in cases with open fontanelles to collect CSF directly through the head from an enlarged ventricle

- **Isotopic cisternography**
  - Radioisotopes injected into ventricles and absorption monitored over days
Treatment
Medical, Surgical, Complications
Medical Therapy

- **Acetazolamide**
  - Carbonic anhydrase inhibitor (diuretic)
  - Decreases CSF production
  - 10mg/kg TID then tapered to effect

- **Diuretics**
  - Frusemide at 1mg/kg SID
  - Reduces CSF production by Na/K/Cl co-transport inhibition
  - Short lived efficacy

- **Omeprazole**
  - Decreases CSF production in dogs by 26% reportedly

- **Prednisolone**
  - 0.25-0.5mg/kg BID

- **Anti-seizure meds as required**
Surgery - Shunts

- Attempt to treat cause first
- Vetriculoperitoneal shunt most common but also ventriculoatrial (right atrium) and lumboperitoneal.

- 3-piece design:
  - Ventricular catheter
  - Valve
  - Peritoneal catheter
Surgery - Shunts

- Ventricular catheter
  - Barium-impregnated silicone
  - Not too stiff, not too compliant
  - Some have a reservoir and port for CSF collection

- Peritoneal catheter
  - Long with many fenestrations

- Most come as separate components which are connected intra-operatively
Surgery - Shunts

Valves

- Most have differential pressure control
- Defined by opening and closing pressures
  - Eg. low pressure (<1 cmH2O), medium (4-8 cmH2O), high (>8 cm)
  - Choice is largely arbitrarily decided but in most vet cases ventricles are very large and so over-drainage complications are a problem – therefore err on the side of higher pressure drain

- Medtronics Strata® – non-invasively adjustable
  - Magnetically adjusted
  - ‘Locked’ percutaneously

- Siphon regulator
  - Prevents rapid over-drainage (discussed later)
Surgery - Shunts

- **Indications**
  - Clear guidelines not established in literature
  - Enlarged ventricles alone is not an indication
  - Clinical signs that worsen despite medical therapy
  - Success depends on degree of cortical loss
    - Need to perform before this occurs

- **Contraindications**
  - Infection, infection, infection….
  - Hydrocephalus ex vacuo
  - Non-recoverable brain disease
Largely the same technique for all types

Asepsis and haemostasis are critical
  - Bipolar electrosurgical cautery
  - Keep shunt packaged until required
  - Don't contact shunt with drapes, gauze, blood

Prep head to abdomen
Directed rostrally from the parietal bone region

Aim to have tip in the frontal horn of lateral ventricle

Rostral or caudal to choroid (ie. not in it)

Required length measured pre-op

Cranial incision 2-3cm lateral to nuchal crest

Abdominal incision just caudal to 13th rib half way dorsoventrally
Surgery - Technique

- **Abdominal incision**
  - Start with just skin and SC incision

- **Cranial incision**
  - Incise skin, SC and superficial muscles
  - Temporalis fascia may be incised to elevate temporalis muscle if preferred/required

- **Tunnelling**
  - Shunt passer (Medtronics™ kit comes with one)
  - Cranial to caudal preferred
  - Pass catheter directly from sterile tray and leave in tray until required
Calvarium entry
- Parietal bone
- High speed burr hole – slightly larger than cath/anchor
- Suture holes can be created adjacent if preferred
- Non-absorbable pre-placed if suture holes used

Dural entry
- Incise sharply and coag with bipolar if required
- 22-g spinal needle through cerebral mantle to assess depth and angle of insertion (collect CSF) then remove
- Bipolar coag pia mater if required
Surgery - Technique

- Catheter placement
  - Push catheter through mantle to determined depth
  - Observe CSF flow
  - Anchor (provided clips/’elbows’ or finger trap suture)

- Attach valve
  - Catheters sutured to valve
  - Valve placed SC against skull more caudally (MR/CT artefact)
Abdominal catheter

- Once attached observe CSF flow distally
- Aim to push at least 1/3 of catheter length into abdomen
- Some catheters have valves on the end so can’t be cut
- Allow excess tubing in the tunnel to accommodate patient movement
- Finger trap suture to abdominal muscle
Complications

- **Blockage**
  - Ventricular end
    - Choroid plexus obstruction (see top image)
    - Glial tissue from astrocyte proliferation
  - Debris in the valve
  - Peritoneal end
    - Scarring or adhesion

- **Over-drainage**
  - Very large ventricles collapse rapidly
  - Cerebral collapse and vascular tearing leads to subdural haematoma/seroma

- **Misplacement, Migration, Disconnection**
  - Technique errors

- **Infection**
  - CSF collection from shunt reservoir
Outcomes & Prognosis

- **Outcome of ventriculoperitoneal shunt implantation for treatment of congenital internal hydrocephalus in dogs and cats: 36 cases (2001-2009), Biel et al, JAVMA 2013**

- 30 dogs and 6 cats – all congenital cases
- 26 (72%) had improved signs following shunting
- 7 dogs and 2 cats (25%) were completely free of signs
- 8 (22%) developed post-op complications
  - 4 occluded (11% of all patients) (2 of these infected also)
  - 3 infected (8.5%)
  - 1 peracute post-op death from over-drainage (2.8%)
  - 1 developed seizures 10 days post-op (2.8%)
  - 1 disconnected catheter (2.8%)
- 55% were alive 18 months post-op
Outcomes & Prognosis

- **Seizures**
  - Biel reported 12/36 (33%) presented with seizures which is comparable to rate in children
  - Biel reported 10/12 dogs were ‘cured’ of seizures although 4 still required meds
  - De Stefani *(VetSurg 2011)* reported a higher rate of seizures (8/12 = 66%) pre-op – suspected to be due to inclusion of acquired cases

- **Blindness**
  - Biel reported 21/36 presented with post-tectal blindness but only 4/21 had vision restored post-surgery
Biel reported gait abnormalities, obtundation and behavioural changes were signs that improved significantly.

Bungee jumping with a shunt??


Levine, *Intracranial pressure and ventricular expansion in hydrocephalus: have we been asking the wrong question?*, J Neurol Sci 2008


