Hydrocephalus:
Ask the Expert

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CSF Formation

- CSF production constant 0.3 cc/minute (18 cc per hour, ~ 400 cc per day)
  - 80% of CSF produced by choroid plexus
  - 20% from ‘extrachoroidal’ sources
  - Active (Na$^+$ pumping) mechanism

- Intraventricular circulation of CSF
  - Foramen of Monro
  - Sylvian Aqueduct
  - Fourth ventricular outlet foraminae
    - Luschka (Lateral), Magendie (Midline)
CSF Reabsorption

- CSF flow through subarachnoid spaces
- Absorption at parasagittal dura (infants), arachnoid granulations (older children/adults)
  - Dural sinuses ---> jugular vein ---> vena cava
  - Probably passive bulk flow
- Accessory drainage sites possible
  - Cervical lymphatics
  - Nerve root sheaths
CSF Reabsorption
Pathophysiology of Hydrocephalus

- Represents imbalance of CSF production and absorption
  - “Bathtub” model
- Majority of hydrocephalus is due to obstruction or inadequate absorption
- Overproduction very rare
  - Choroid plexus tumors
  - Villous hypertrophy of choroid plexus
Hydrocephalus: Traditional Classification

- **Obstructive (IVOH)**
  - **Congenital**
    - Aqueductal Obstruction
    - Atresia of Monro
    - Chiari malformation
    - Dandy-Walker
    - Intracranial cysts
  - **Acquired**
    - Infectious
    - IVH
    - Chemical/Adhesive
    - Neoplasm, cyst, mass

- **Communicating (EVOH)**
  - **Congenital**
    - Chiari Malformation
    - Dandy-Walker
    - Incompetent arachnoid villi
    - Encephalocele
    - Intracranial cysts
  - **Acquired**
    - Infectious
    - SAH
    - Chemical/Inflammatory
When To Treat?

- **Expectant Observation**
  - Macrocraania and enlarged extra-axial spaces ± mild ventricular enlargement
    - Asymptomatic
    - Normal developmental milestones
    - Normal fundoscopic examination
- **Clearcut Indications for Treatment**
  - Very large or rapidly enlarging ventricles
  - Clear-cut symptoms
When To Treat?

- Child with moderately enlarged ventricles
  - Assess head growth, fontanelle, sutures, etc.
  - What degree of ventricular enlargement interferes with normal development?
  - How does the risk of ‘compensated hydrocephalus’ compare with the risk of shunt complications?
When To Treat?

Young et al., Pediatrics 52:38 (1973)

- **Post-shunt** cortical mantle at least 2.8 cm
  - Normal IQ
- Post-shunt cortical mantles < 2.0 cm
  - Subnormal IQ
- No differences in IQ for children 2.0 - 2.8 cm
- Cortical mantle > 2.8 reached in children shunted by 5 months of age
- Supports observation if cortical mantle > 2.8 cm
- Expectant observation for 2.0-2.8 cm??
When To Treat?

Four children with ‘compensated’ hydrocephalus had improved intellectual performance following delayed shunting at 4.5 – 8.5 years of age.

- Improvements in neuropsychometric testing, IQ
- No consistent improvement in achievement scores

Criticisms: Not a representative population

- 3 of 4 had focal neurological deficits, prior insult
- 1 of 4 had prior trauma with subdural hematomas

Torkelson et al., J Neurol, Neuros, Psych 48:799 (1985)
Survey of Pediatric Neurosurgeons

Attendees at AANS/CNS Pediatric Neurosurgery meeting, Spokane, 2008

- Provided with 22 clinical scenarios
- Given sheet with 5 sets of CT scans showing progressive ventricular enlargement
- Asked to indicate *minimum* ventricular size they would shunt for each scenario
6 wk asymptomatic infant
9 mo, normal development and normal extra-axial spaces.
9 mo, normal development, widened EAS, FH of macrocephaly
7 yr, concussion, normal 2nd grader
7 yr, concussion, struggling in 2nd grade
Conclusions

- Pediatric neurosurgeons are relatively ‘conservative’ in recommending surgery for hydrocephalus in asymptomatic children
  - Especially so if head circumference tracking near 50%ile and paralleling curve
- Rarely (< 10% of respondents) shunt mild-moderate sized ventricles
Conclusions

- Majority (> 50% of respondents) would not shunt even largest ventricles presented in face of HC curve stable at 50th percentile
- More likely to shunt normal infants/toddlers than normal 7 y/o with accelerating HC
- Less likely to shunt 9 mo old in face of enlarged EAS and + FH of macrocephaly
- More likely to shunt in face of developmental delays in all scenarios
Treatment Options

- Shunts
- Endoscopy
- Choroid plexus cauterization
There are only three types of shunts:

- Blocked shunts,
- Infected shunts,

and blocked and infected shunts”

E. Bruce Hendrick

Hospital for Sick Children

Toronto
Shunt Complications

- **Shunt Failure**
  - 30-40% within first year
  - 50 - 60% at 2 years
  - 85% at 10-12 years
  - Revision takes failure rate back to 1st year again

- **Most common sites of shunt failure**
  - Ventricular catheter obstruction with debris, choroid plexus
  - Disconnection, fracture (primarily older shunts)
  - Valve obstruction
  - Distal tubing
Shunt Survival Rates
Shunt Failure - CT Scan

Baseline Study

Shunt Failure
Case 1

7 year old boy with shunted hydrocephalus
- Headaches for 4 days, 6/10
- Intermittent, makes him cry, awakens at night
- No emesis, no fevers, infectious symptoms, or sick contacts, no personal or FH of migraines
- No papilledema
- Neurological and general examination normal
- Shunt appears intact
Case 1

- CT scan at presentation
Case 1

CT at baseline

CT at presentation
Case 1

- Need prior scans to compare
- Approximately 10-15% of children with little or no increase in ventricular size
- History and Examination are important
  - Making him cry, stop playing
  - Awakening from sleep, present before getting OOB
  - Lack of infectious symptoms or sick contacts
  - Similar to other shunt headaches
  - Parents worried this is shunt related
  - Papilledema (always check eyes)
Case 2

- 9 year old with spina bifida and shunted hydrocephalus
  - Straight caths, usually dry between caths
  - Increased urinary incontinence past 3 months
  - Urinalysis benign, culture negative
  - Further questioning reveals intermittent back pain at spina bifida closure site, but no headaches
  - Neuro examination unchanged, no papilledema
Case 2

- Spina bifida presents unique challenges
- Virtually any deterioration may be shunt related
  - Headaches, nausea, vomiting, neck pain
  - Seizures (rare in isolation)
  - Decline in school performance, cognitive function
  - Worsening swallowing dysfunction, aspiration, apnea
  - Back pain (esp at closure site), scoliosis
  - Decline in sensorimotor function (arms or legs)
  - Decline in urinary or bowel function
- **Always check the shunt first !!!**
Case 3

- 15 year old with prior history of brain tumor and shunted hydrocephalus, presents with postural headaches 2 days after routine LP for CSF cytology
  - Headaches clearly worse when upright, better when recumbent
  - Nausea, emesis when sitting > 5 minutes
  - Listless, no fever, nuchal rigidity, or neuro deficits
Case 3

- CT scan at presentation
Case 3

- Shunt tap performed
  - Opening pressure < 0 cm H$_2$O
  - Can easily aspirate 10 cc CSF from shunt
  - Patient worse after tap
Case 3

- Shunt tap performed
  - Opening pressure < 0 cm H₂O
  - Can easily aspirate 10 cc CSF from shunt
  - Patient worse after tap
- Patient placed at strict bedrest for 48 hours
- CT scan repeated 2 days later, ventricles back to baseline, patient now asymptomatic
- Arises now without difficulty, no recurrence of symptoms
Case 3

- Patient has a relative degree of **intraventricular obstructive hydrocephalus**
  - Ventricular CSF cannot communicate readily with CSF in subarachnoid space or spinal theca
- Post-LP produces a low pressure syndrome which maintains a **low global intracranial pressure**
- Intraventricular pressures never exceed valve pop-off pressure setting
- Ventricular CSF can’t escape and accumulates under low pressure
12 year old with shunted hydrocephalus, last revised 5 years ago

- Presents with 5 mo history of slowly progressive abdominal distension and mild, dull abdominal pain
- No headaches, vomiting, fevers, hematemesis, melena, hematochezia, constipation or diarrhea
- Examination discloses abdominal distension with large, soft and nontender RLQ abdominal mass
- Rectal exam normal, stool heme negative
Case 4

- Abdominal ultrasound demonstrates a large fluid filled mass in RLQ, surrounding shunt catheter tip
Abdominal ultrasound demonstrates a large fluid filled mass in RLQ, surrounding shunt catheter tip

- Tap 1500 cc of fluid with slight xanthochromia
  - 38 WBC, 2 RBC, 450 protein, 84 glucose
  - Gm stain and cultures negative after 3 days
Case 4

- Abdominal ultrasound demonstrates a large fluid filled mass in RLQ, surrounding shunt catheter tip
- Tap 1500 cc of fluid with slight xanthochromia
  - 38 WBC, 2 RBC, 450 protein, 84 glucose
  - Gm stain and cultures negative after 3 days
- Anaerobic cultures of fluid grow P. Acnes
  - Shunt removed, treated with PCN for 7 days, new shunt reinserted
Case 4

- **Shunt Infections**
  - 5-8 % overall
  - Most frequent (70%) within 8 weeks post-op
  - 80 % within first 6 months post-op
  - Staph epi > Staph aureus > P. Acnes > GNR

- **Most common presentation is SHUNT FAILURE**
  - Headaches, anorexia, nausea, vomiting
  - Fever (usually low grade), meningismus (25-33%)
  - Erythema/swelling along tract
  - Abdominal pain, pseudocyst
Diagnosis made by shunt tap

- Preferably done before antibiotics are begun!!!!!
- Lumbar puncture inadequate (50% sensitivity) and may be dangerous (herniation)
- WBC and protein increased, glucose decreased, but may be unremarkable!
- Gram’s stain may not be positive with S. Epi, P. Acnes
- Cultures usually positive within 72 hours
  - Exception: P. Acnes (7 days, anaerobic cultures)
Shunt Infection - Treatment

- **Remove shunt**
  - Insert temporizing external drain (EVD)
  - Serial CSF cultures until negative

- **Treat with broad spectrum antibiotics**
  - Vancomycin, third generation cephalosporin
  - Rifampin for intracellular staph organisms
  - Adjust antibiotics after sensitivities known
  - 7-10 days

- Re-insert new shunt hardware
Alternative Treatments

- Full course of antibiotics followed by removal and replacement of shunt
  - Requires only 1 operation
  - Less frequently effective (~80-85%)
- Antibiotic treatment without shunt removal
  - Effective only for S. Pneumo, H. Flu
    - Hematogenous spread to shunt
    - Combined intravenous and intraventricular antibiotics for S. Epi (McLaurin method)
      - Effective in only 60%
Case 5

- 4 year old with shunted hydrocephalus due to aqueductal stenosis in infancy, presents with 2 day history of severe headaches, emesis
  - Parents note ‘eyes are funny, don’t move together’
  - ‘Never been like this before’
  - Listless, papilledema, mild VI nerve palsy
  - No other focal neurological abnormalities
Case 5
Endoscopic Third Ventriculostomy (ETV)

- Option for patients with IVOH
  - 2 yrs or older
  - 80% success rate

- Spina bifida
  - Anatomy difficult
  - 60-70% success rate

- Other forms of HC
  - 30-50% success rate
Endoscopy

- ETV
- Septal fenestration
- Fenestration of intraventricular adhesions
- Placement of shunt catheters
- Removal of intraventricular tumors, cysts
- Endoscopic assistance for open approaches
- ETV plus endoscopic choroid plexus cauterity
Hydrocephalus: Conclusions

- Listen to parents!!! History important
- Characteristics, circumstances of headaches
- Need baseline CT scan, CT may change little or not at all in some children
- May need admission for observation in questionable cases
- *Keep an open mind* - if child is worsening, find a neurosurgeon who will listen and act
- It’s not always just plumbing!