Hydrocephalus: Where Have We Been? And Where Are We Going?

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Objectives

1. The participants will be able to identify the different types of hydrocephalus
2. The participants will be able to recognize the different treatment modalities of hydrocephalus
3. The participants will be able to identify the signs and symptoms of hydrocephalus
4. The participants will recognize the diagnostic criteria for Normal Pressure Hydrocephalus.
Hydrocephalus: “Water on Brain”
- “hīdrōˈsefələs”
- Hudrokephalon (Greek)
- Dysfunctional cerebrospinal fluid
- AKA:
  - Hydro
  - HCP
Hydrocephalus

Cerebrospinal Fluid

Surrounds the spinal cord
Fills ventricles within the brain
Blood-brain barrier controls which solutes enter the cerebrospinal fluid
Ventricles

Ventricular System:

• Lateral Ventricles (2)
  – Foramen of Munro

• 3\textsuperscript{rd} Ventricle
  – Aqueduct of Sylvius

• 4\textsuperscript{th} Ventricle
  – Foramen of Lushka (2)
    • Lateral
  – Foramen of Magendi
    • Midline
Choroid Plexus
Cerebrospinal Fluid

- Produced by choroid plexus
- Absorbed by arachnoid granules
- ~140ml total CSF
- Rate of 0.2-0.7ml/kg/hr
  - 25-30ml/hr
  - 600-700ml/day
- Relies on Na+/K+ ATP dependent pump and carbonic anhydrase
- Acetazolamide can partially decrease production of CSF
  - Can cause hypercapnia that can increase CBF and possibly ICP
- Other inhibitory agents:
  - Halothane
  - Angiotensin II
  - Vasopressin (V1 receptor)

NO OFF SWITCH!
Hydrocephalus
Hydrocephalus

Categorizations:

• CSF dynamics
• Timing
• Etiology
• Intracranial Pressure
• Age of patient
CSF Dynamics

• Obstructive Hydrocephalus
  – Aqueductal Stenosis
  – Cysts (colloid cysts, neuroenteric cysts)
  – Tumors (cerebellar tumors, intraventricular tumors)

• Communicating Hydrocephalus
  – AKA “non-obstructive hydrocephalus”
  – Infection (meningitis)
  – Hemorrhage (subarachnoid)
  – Age (normal pressure HCP).
Hydrocephalus Timing

• Acute Hydrocephalus
  - Life threatening
  - Due to hemorrhage, tumors

• Chronic Hydrocephalus
  - Compensated by brain
  - Delayed outcome following:
    • Infection
    • Hemorrhage
    • Congenital malformation

• Normal Pressure Hydrocephalus

Chronic

Acute
Elevated ICP

Signs and Symptoms:
1. Headache
2. Nausea/Vomitting
3. Blurred Vision / Diplopia
4. Focal Neurological Findings
   1. Dilated pupil(s)
   2. Hemiparesis
5. Altered Mental Status
   1. Confusion
   2. Lethargy
   3. Obtundation
   4. Stupor
   5. Coma

Cushing’s Reflex:
- Hypertension
- Bradycardia
- Irregular Respirations
Monroe – Kellie Hypothesis

Monroe - Kellie Hypothesis:
(Not really a doctrine...)

1. Skull is a closed box
2. In the setting of a mass lesion:
   1. Displaceable fluids will compensate first
   2. Elevated intracranial pressure subsequently occurs
Unique findings with hydrocephalus:

- Headaches (worse when lying flat)
- Papilledema
- Diplopia (Horizontal)
- Parinaud's Syndrome
Parinaud’s Syndrome

Due to lesion compressing “tectal plate”

1. Upgaze paralysis
2. Argyll-Robertson Pupils
   1. (light-near dissociation)
3. Convergence-retraction nystagmus
   1. With upgaze or opticokinetic drum spinning down
4. Eyelid Retraction (Collier’s Sign)
5. Conjugate down-gaze (sun-setting sign)
Other CSF syndromes

• Normal Pressure Hydrocephalus
  – Elderly population
  – Treatable cause of dementia

• Pseudotumor Cerebri
  – Symptoms of hydrocephalus (or tumor)
  – Normal CT or MRI
  – Elevated ICP (on LP)
  – One etiology is obesity

• “Slit Ventricle Syndrome”
  – In Chronically overshunted patients
  – Stiff ventricles (no change across wide pressure)
  – CT Scans look normal or overdrained (slits)
HCP Management

• Acute or Not?
  – Should we wake up the OR team?
  – Can we wait for the OR?
• Etiology
  – Tumor
  – Hemorrhage
• Patient status
  – Hemodynamically unstable (Cushing’s response)
  – Infection / Meningitis
HCP Management

1. Elevation of Head > 30°
2. Hyperventilation
3. Diuresis
4. Sedation
5. Metabolic Coma
6. Systemic Hypothermia
7. Ventriculostomy
8. Surgery
Ventriculostomy

- Bedside Procedure
  - 15 minutes to place
- Rapid reduction in ICP
- Can directly measure ICP and treat by CSF drainage

Caveats:
- 30% hemorrhage rate (CT)
- 4% daily infection rate
- “Blind procedure”
- Can be dislodged
  - (patient / nurse / tech / family / pet, etc...)
Medical Management

• NOT first line for Acute Hydrocephalus!

• acetazolamide (Diamox)
  – Carbonic Anhydrase inhibitor
  – Temporary effect (equilibrates in 3-4 days)
  – Can cause respiratory failure at high doses

• Mannitol
  – Sugar Acid
  – Rapid decrease in ICP
  – Lasts 3-4 hours

• Volatile Anesthetics
  – Halothane
  – Isoflurane

• Vasopressin / Angiotensin II
Surgery for HCP

Internal CSF Diversion
- Ventriculoperitoneal Shunt (VP Shunt)
- Ventriculopleural Shunt (VPL Shunt)
- Ventriculoatrial Shunt (VA Shunt)

Internal CSF Bypass
- Endoscopic Third Ventriculostomy (ETV)
  - Bypass created in floor of 3rd Ventricle
- Torkalson Shunt
  - Implanted bypass between Lateral ventricle and 4th Ventricle
  - Not frequently used
Shunts

VP Shunt

- Most common type in adults
- Peritoneum can absorb liters of fluid daily
- (Total daily production of CSF ~500cc)
- Valve separates ventricular catheter and peritoneal catheter
  - Fixed or Programmable
Ventriculopleural Shunt

- More common type in children
- Pleural space more limited
- Associated with pleural effusions
Ventriculoatrial Shunt

- “Backup” site if other sites fail
- No limit to CSF absorption
- Helpful for difficult to drain patients (low pressure hydrocephalus)
Shunts

Lumboperitoneal Shunt

• Specific indications:
  • Pseudotumor Cerebri
  • Chronic CSF rhinorrhea
    – Post-traumatic
    – Iatrogenic
  • High Failure Rate
# Shunts

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<thead>
<tr>
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<th>VP Shunt</th>
<th>VA Shunt</th>
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<tbody>
<tr>
<td><strong>Pros</strong></td>
<td>Easier to place</td>
<td>Avoids Abdomen</td>
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<tr>
<td></td>
<td>Less chance of overdrainage</td>
<td>Low Resistance (negative pressure)</td>
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<tr>
<td></td>
<td></td>
<td>Easily Convertible</td>
</tr>
<tr>
<td><strong>Cons</strong></td>
<td>Abdominal organ injury</td>
<td>More difficult to place</td>
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<tr>
<td></td>
<td>Peritonitis necessitates removal</td>
<td>Overdrainage a concern</td>
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<td>Higher back-pressure (10mmHg)</td>
<td>Risk of stroke during placement</td>
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Shunt Valves

Fixed
• Distal Slit
• Fixed pressure intervening valve

Programmable
• Large Range
• High Resolution
• High Pressure

MRI-Lock
• Resistant to MRI reprogramming
For the adjustment: position is unlocked with a specific magnetic key
Shunt Woes

• Hydrocephalus: Complicated Disorder
• VP Shunt: Complex Hardware (not a simple device)
• Numerous Challenges for Caregivers

1. Failure: ~10% / year (asymptotic curve)
   – Some reports upwards of 50% failure in 1st two years
   – Tube kinks / separations
   – Valve obstruction

2. Infection: 1.6-16.7%
   – Highest risk in 1st month after surgery
   – Multiple revisions major risk factor

3. Over-/ Under-drainage
Endoscopic Third Ventriculostomy

Ideal for Obstructive Hydrocephalus

- Requires relative pressure gradient for patency
- Auto-equilibrates to prevent overdrainage complications
- No hardware in brain
- Can still have failure rate (~20%)
  - Missed in ED as patients are unaware or unconscious and caregiver does not know to evaluate for HCP
Endoscopic Third Ventriculostomy
Endoscopic Colloid Cyst Resection
# Shunt v ETV

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<tr>
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<th>Shunt</th>
<th>ETV</th>
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<tbody>
<tr>
<td><strong>Pros</strong></td>
<td>More Common Procedure</td>
<td>No Hardware</td>
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<tr>
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<td>Adjustable Pressure / Flow</td>
<td>Little risk of Overdrainage</td>
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<td>Dependent on Technology</td>
<td>Dependent on Technology</td>
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<tr>
<td><strong>Cons</strong></td>
<td>Failure Rate (10% / year)</td>
<td>Failure Rate (2% / year)</td>
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<tr>
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<td>Obstructive Hydrocephalus</td>
<td>Post-infectious (basilar meningitis)</td>
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<td>Abdominal Organ Injury (or cardiac valve injury)</td>
<td>Basilar Tip Aneurysm</td>
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Normal Pressure Hydrocephalus

Clinical Triad:
1. Magnetic (shuffling) Gait
2. Urinary Incontinence
3. Dementia

... In the setting of enlarged ventricles

- One of very few *Treatable* causes of dementia!
- Prevalence: 21.9/100,000 patients
- 3.3/100,000 (50-59yrs)
- 49.3/100,000 (60-69yrs)
- 181.7/100,000 (70-79yrs)
- 1% of all dementia patients
Normal Pressure Hydrocephalus

Diagnosis:

1. Clinical:
   - Assess Gait, speed and quality
   - Assess memory

2. Radiographic:
   - Evan’s Index >0.3

Dynamic Testing

1. High Volume Lumbar Puncture
   - 20-40ml (depending on patient)

2. Lumbar Drain Trial
   - Hourly drainage of 10-15ml CSF
   - Over 3 days (in ICU)
   - Daily assessments of MMSE and PT
   - 84% of responders had successful VP Shunt outcomes (90% PPV)
     - Marmarou et.al. JNS 2005
Normal Pressure Hydrocephalus

After Surgery...

- Generally well tolerated operation with low major risks
- Some studies suggest upwards of 38% overall complication rate
- Need to monitor patients closely and identify target drainage pressure
- Avoid aggressive adjustments to prevent subdural hematomas
- Incremental and regimented adjustment of VP Shunt settings helpful for objective outcomes assessment
After Surgery:

• Generally well tolerated operation with low major risks
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Future Directions

VP Shunts are artificial solutions to underlying pathology

• Overproduction of CSF
• Underabsorption of CSF
• Obstruction of CSF flow

Ideal goal – eliminate need for VP Shunt altogether...!

• Choroid Plexus Coagulation (CPC)
Future Directions

- CPC + ETV 66% efficacious v ETV alone (47%)
- Most etiology demonstrates improved outcomes

- Other studies showed positive results in older children
- Potentially can apply to adults...
www.brain-tumor.org
John Wayne Cancer Institute