Severe Headaches in Patients with shunts
Slit Ventricle Syndromes
Incidence Of Headaches In Shunted Hydrocephalus

- Essentially all adolescents and young adults with shunts will have headaches if asked
- Most of these headaches are mild, intermittent and lead to normal function
- Severe headaches in shunted patients with apparently working shunts is called the “Slit Ventricle Syndrome” (SVS)
Slit Ventricles

- Radiographic slit ventricles – 80%
- Slit Ventricle Syndrome – 15%
- Without symptoms there is no need to treat
Causes Of Headaches Based On ICP Monitoring

• Intracranial Hypotension
• Intermittent proximal obstruction
• Intracranial hypertension with a failed shunt and small ventricles (Normal Volume Hydrocephalus)
• Intracranial hypertension with a working shunt (Cephalocranial dysproportion)
• Shunt Related Migraine
Headache Syndromes

- Intense headache: 10-90 minutes
- Headaches worsening in afternoon: better lying down
- Awakens patient from sleep
- Positive family history
- Intermittent proximal obstruction
- Intracranial hypotension
- NVH or CCD
- Shunt related migraine
Medical Management Of Headaches: Patients with Shunts

• Similar to with migraine
• Beta blocker: Inderal
• Cyproheptadine
• Sumatryptin (Imitrex) and analogs
• strategies
Who Needs Intervention

- Patient should decide when and how to intervene
- Suggested threshold: Must leave school or work twice per month or more frequently
- Lying down in school nurse’s office
- Most of these patients are not at imminent risk and decision can be made over time
- The headache diary
The First Step

• Shunt revision with incorporation of a DRS
• 85% of chronic headaches are improved by this technique only
• Technology of shunting has made great strides in the past decade.
• What does nature intend?
What Is A DRS

- Device which retards siphoning
- Multiple types and designs
- Orbis Sigma
- Delta Valve
- Anti-Siphon Device
- Siphon Control Device
- Siphonguard
- Gravity compensating device
Shunt Removal Protocol

- Exteriorize the distal end of the shunt or remove and replace it with an EVD
- Raise drainage height or clamp ventriculostomy in ICU setting
- Obtain imaging study
Following Imaging Study

Shunt Clamped

Ventricles Enlarge:
- Patient symptomatic
  - Perform ETV

Ventricles Enlarge: Patient asymptomatic
  - Remove Shunt

Ventricles do not enlarge: ICP increased
  - Assess for Lumbo-peritoneal shunt

Normal Volume Hydrocephalus
How to Think
The New Classification
Lumbo-peritoneal Shunt

• Requires valve except in NPH
• Requires Communication be verified
  – Iohexal ventriculogram followed by CT of C-spine
• Value of programmable valve for fine-tuning
• Best with ventricular reservoir left behind
  – The Caleb Procedure
Effect of Blocked Shunt

Brain Volume 1,882 cc

CSAS

Rvent=1.9 cm
Rbr=7.96
Rcr=7.66
Implication Of Non-expanding Ventricles

• Cortical Subarachnoid Spaces (CSAS) must be involved in the hydrocephalic process
• Usually implies venous hypertension is the initial cause of the hydrocephalus
• Does not occur when process begins as adults
• The reasons they are not candidates
  – Can’t manipulate the scope
  – Point of Obstruction is distal to interpeduncular cistern serviced by the ventriculostomy
Assessment And Treatment Of “Normal Volume Hydrocephalus

Ventricles do not enlarge

Perform Iohexal Ventriculography

CT Scan of Head and Cspine

Positive (28/31)
- Valved LP Shunt

Negative
- VP with valve upgrade
- CM to VP
“Normal Volume Hydrocephalus”

• Must Involve the Cortical Subarachnoid Space
• One in 5 children will be found to have non-responding ventricles at the time of shunt failure
• All will have been sent away from ER while still sick
Strategies To Overcome Severe Slit Ventricle Syndrome (NVH)

- Opening pressure (>5mmHg)
- Lumbo-peritoneal shunt (Must have valve)
- Shunt to cisterna magna
- Cisterna magna to ventricle to peritoneal shunt with LP shunt tubing in cisterna magna
- Pressure of valve must be higher than the sagittal sinus
WHAT WORKS

• AVOID SHUNTING IF AT ALL POSSIBLE
• CORTICAL SUBARACHNOID SHUNT TO PERITONEUM
• VENTRICULAR SHUNTS WITH VERY HIGH RESISTANCE
• VALVES IN SERIES
• CISTERNA MAGNA SHUNT TO PERITONEUM
THE CALEB PROCEDURE
Cisterna Magna To Ventricle
To Peritoneal Shunt
Ventricles Enlarge:
Patient Symptomatic

**ETV**
Assess by imaging, ICP and clinically

Shunt Removed: 80%

Failed: consider LP Shunt
Classification of Hydrocephalus: Dandy

• Based on recovery of supra-vital dye
  • Non-communicating hydrocephalus
    – Aqueduct
    – Fourth Ventricle
  • Communicating hydrocephalus
    – Blockage between the SSAS and CSAS
    – Obstruction at arachnoid villi
    – Venous Hypertension
Classification Based On Point Of Obstruction

• All Hydrocephalus is Obstructive
• Ransohoff Classification
  – Intraventricular obstructive
  – Extraventricular obstructive
• Current imaging technology should allow the definition of the “first” point of obstruction
Please Note

• The point of obstruction in post-hemorrhagic hydrocephalus
  – Acutely it is in the area of the arachnoid villi
  – Late, in all forms of PHH the blockage is between the SSAS and CSAS, an ideal case for ETV
What Is Accomplished by Performing an ETV?

- Dural venous sinuses
- Lymphatics
- Nerve root sleeves
- Olfactory bulbs
- Transependymal parenchymal capillaries
Candidates For Ventriculostomy

- Yes
  - Aqueduct
  - Fourth ventricle
  - SSAS to CSAS

- No
  - CSAS to SSS (arachnoid villi)
  - Venous Hypertension (Pseudotumor)
Risk Assessment

• Shunt
  – Infection: 8%
  – Failure: 20-50%
  – Death: 1%/yr

• ETV
  – Hormonal Difficulty usually DI:
  – Loss of recent memory
  – Diplopia
  – Hemiparesis
  – All 3% acute/1% permanent
  – Death
Benefit Assessment

• Shunt
  – It is easy to tell whether or not the hydrocephalus is being treated
  – Initial risks more inconvenient than dangerous

• ETV
  – No reliance on implanted foreign Body
  – Most series show substantially greater longevity
ETV Is Substantially More Dangerous Than A Shunt Procedure and Substantially Less Dangerous Than A Lifetime Of Shunt Dependency
Contraindications to ETV

- Achondroplasia
- Craniofacial Syndromes
- History Of unresponsive ventricles
- Associated with Spina Bifida
  - Controversial
  - Multiple sites of obstruction
  - Danger of assuming shunt independence
Late Outcome

• Personal Experience
  – After two weeks only one patient has failed late
  – That patient failed because of herniation of Basilar artery
  – Late failures try second ETV

• Literature
  – Growing number of late failures
  – Drake et al see ETV failure as same as shunt failure
  – Several Late Deaths
The New Rules

• Every shunt failure is a chance to test shunt dependency
• Shunts are evil
• Most patients with communicating hydrocephalus are candidates for ETV
Conclusions

• At least 70% of shunted patients may be candidates for shunt removal
• Communicating hydrocephalus is a misnomer
• Previously shunted patients are excellent candidates for ETV
• “If it aint broke don’t fix it”

—But

• Make certain that you and your neurosurgeon have a plan for the next step
Rekate’s Rules Of Problematic Shunt Management

• Make certain shunt is really needed
• Attempt shunt removal
• If remains shunt dependent make certain that all CSF compartments see the same pressure either internally or externally
• Make certain ICPs 5-15 mmHg recumbant and –5 to +5 mmHg standing