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?

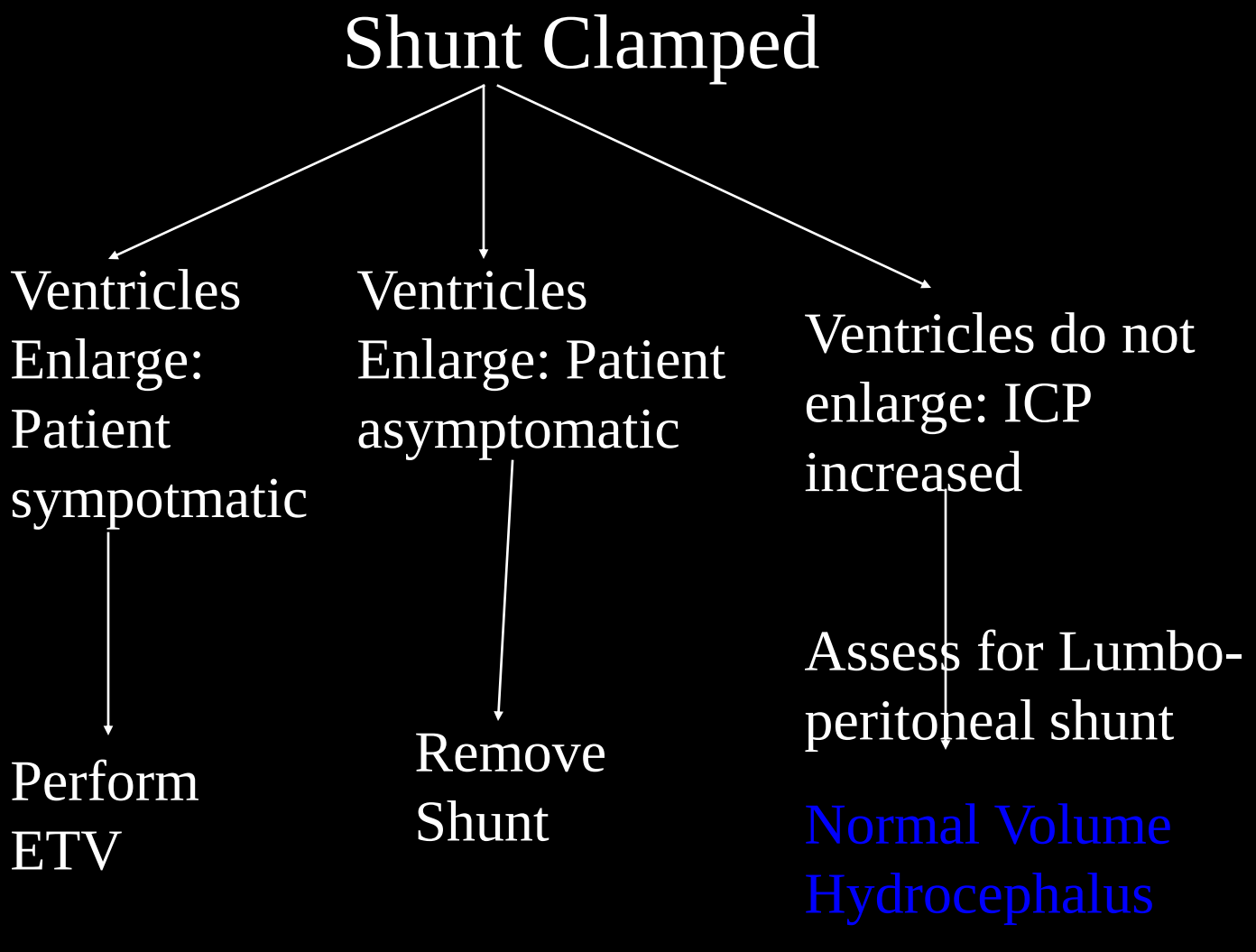
What Is A DRS

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

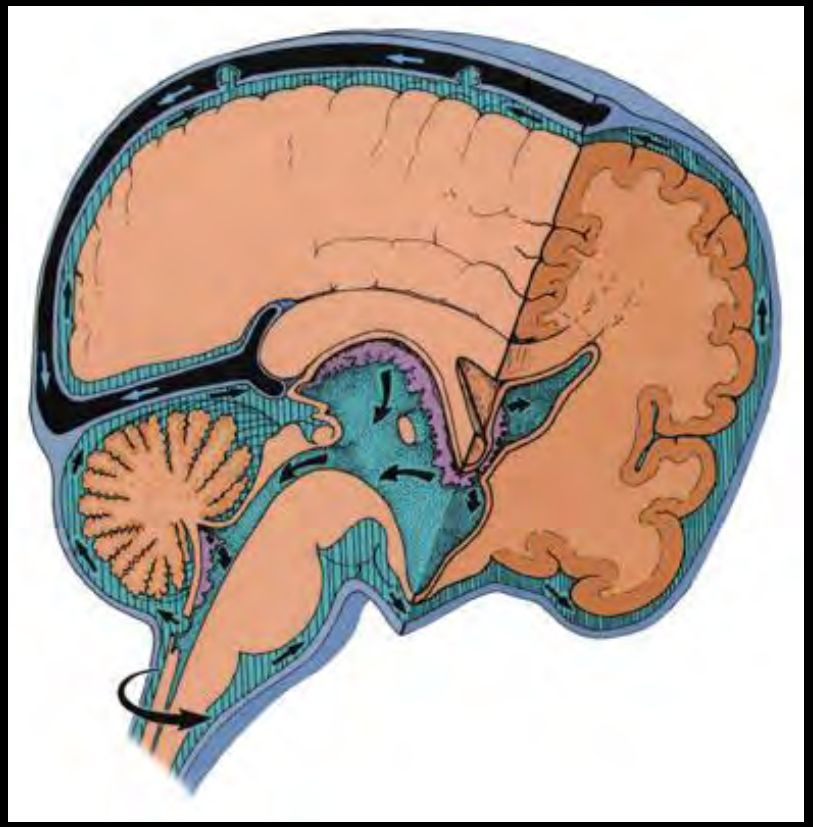
Shunt Removal Protocol

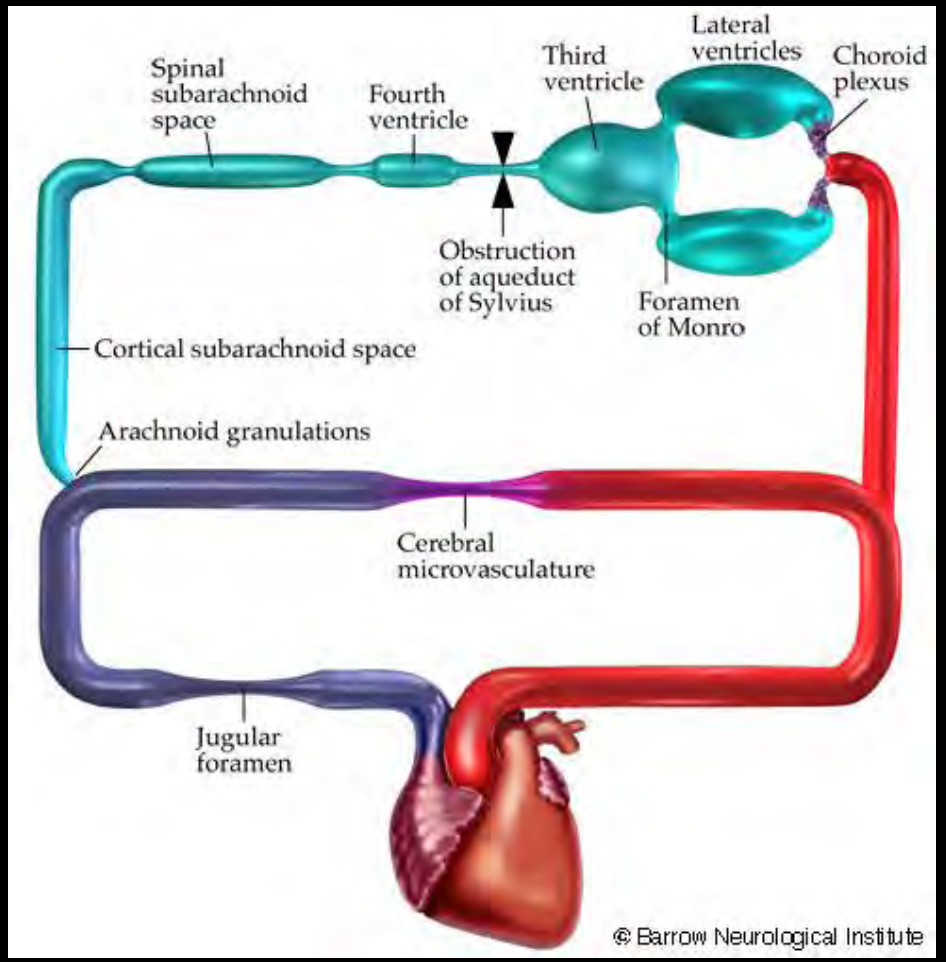
- Exteriorize the distal end of the shunt or remove and replace it with an EVD
- Raise drainage height or clamp ventriculostomy in ICU setting
- Obtain imaging study

Following Imaging Study



How to Think
The New Classification

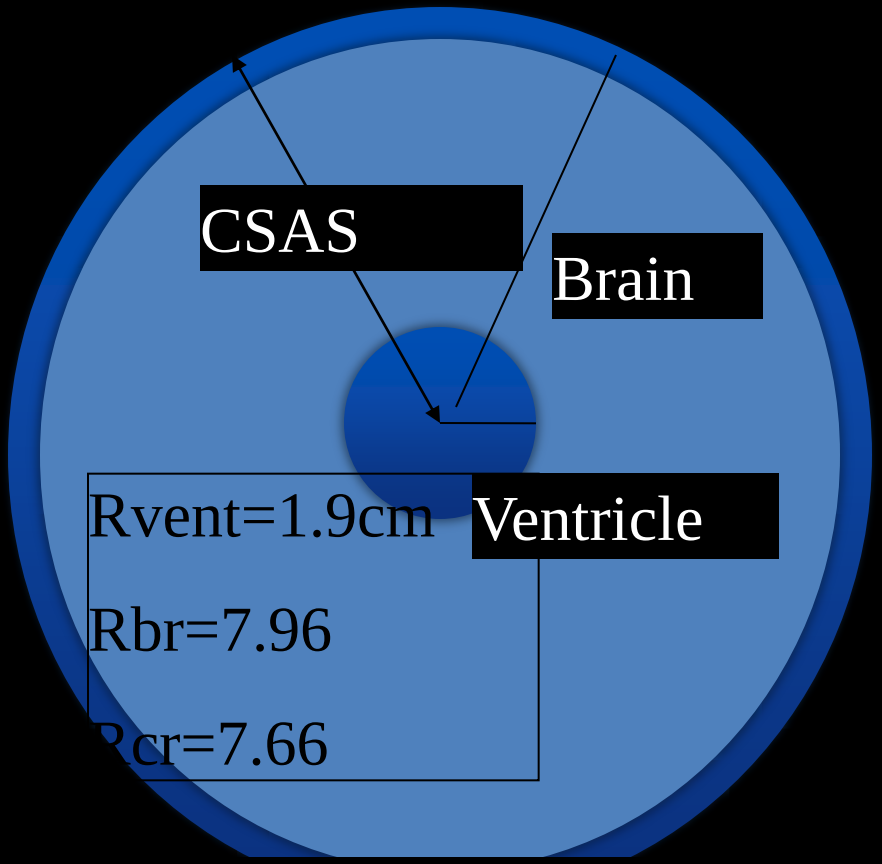




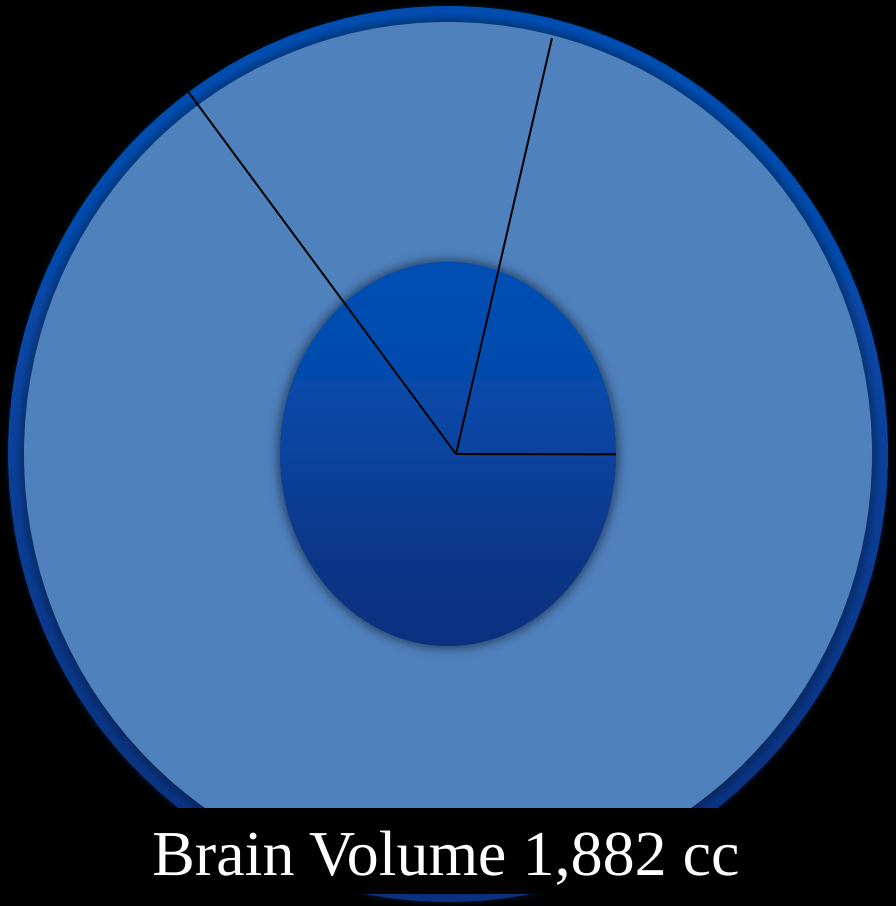
Lumbo-peritoneal Shunt

- 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

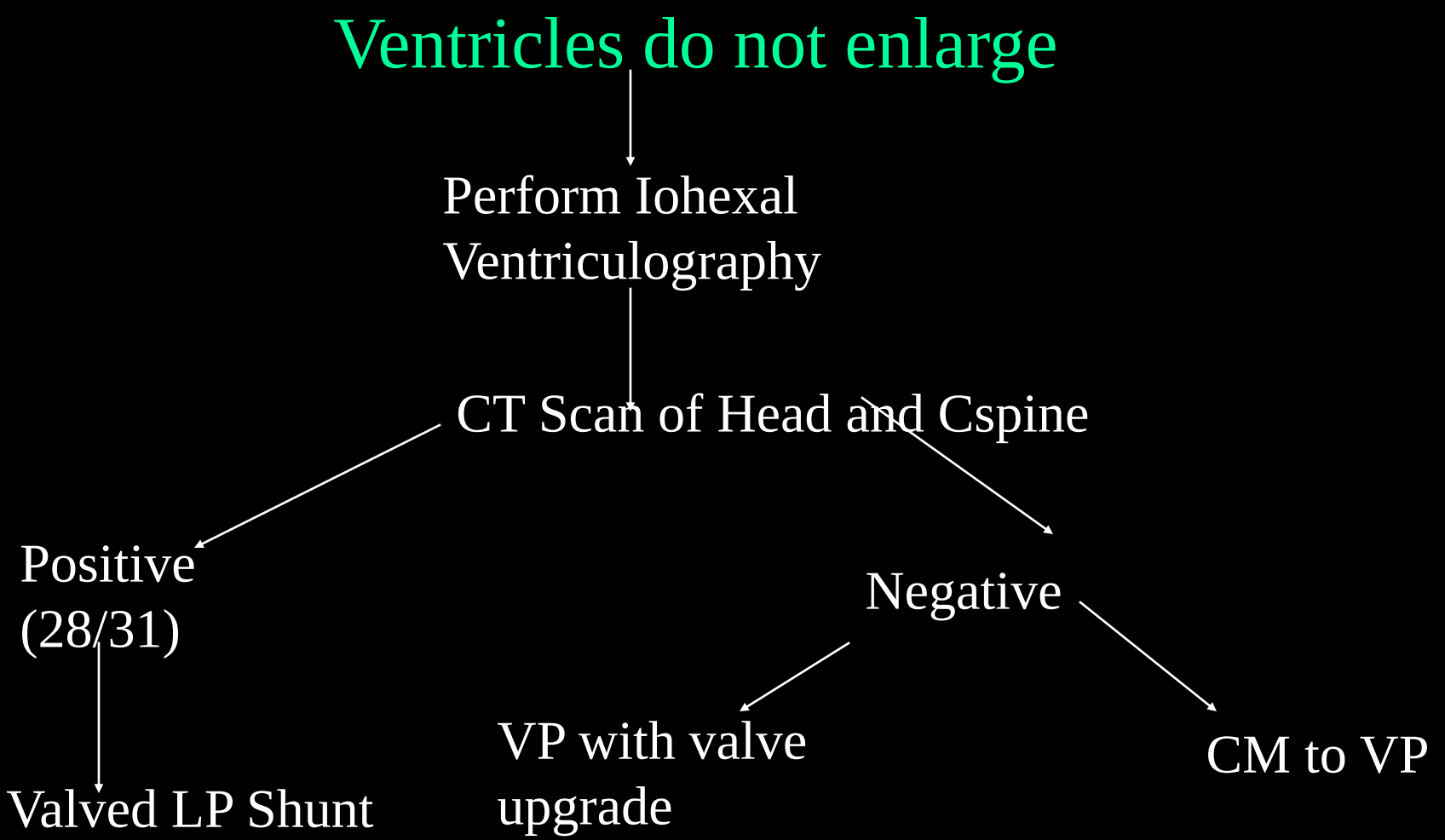


Brain Volume 1,882 cc

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



“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

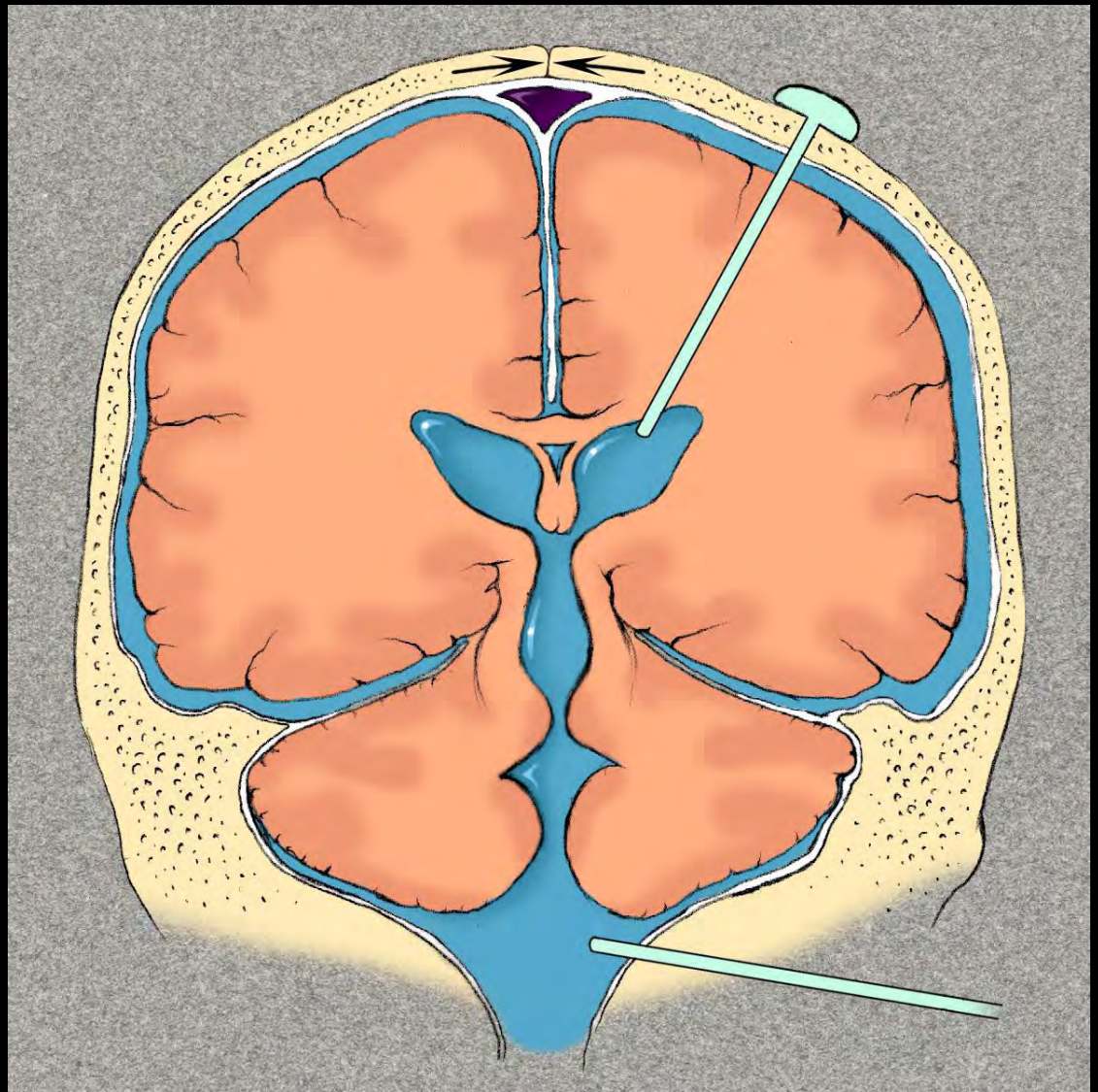
Strategies To Overcome Severe Slit Ventricle Syndrome (NVH)

- 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

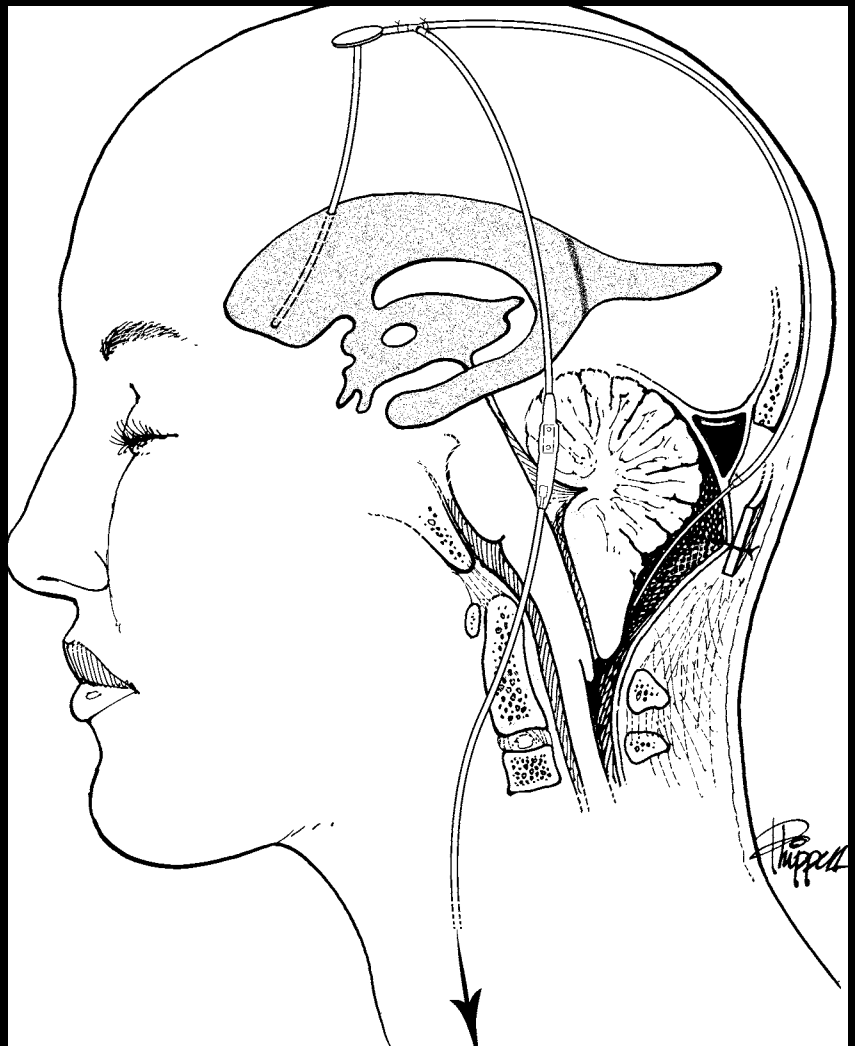
WHAT WORKS

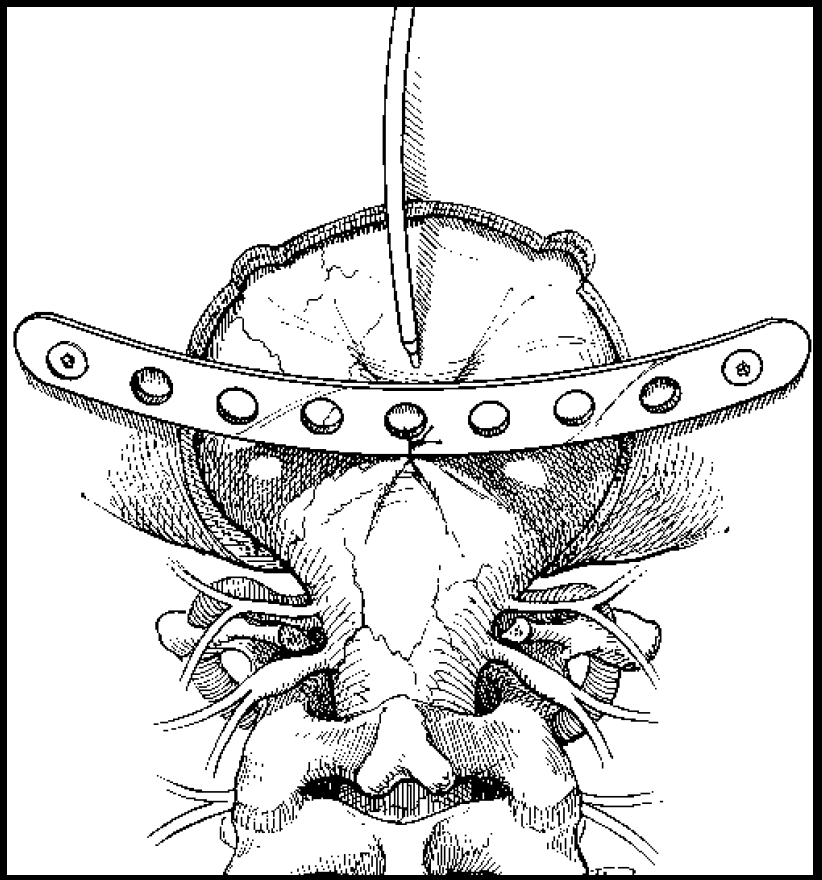
- AVOID SHUNTING IF AT ALL POSSIBLE
- CORTICAL SUBARACHNOID SHUNT TO PERITONEUM
- VENTRICULAR SHUNTS WITH VERY HIGH RESISTANCE
- VALVES IN SERIES
- CISTERNA MAGNA SHUNT TO PERITONEUM

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

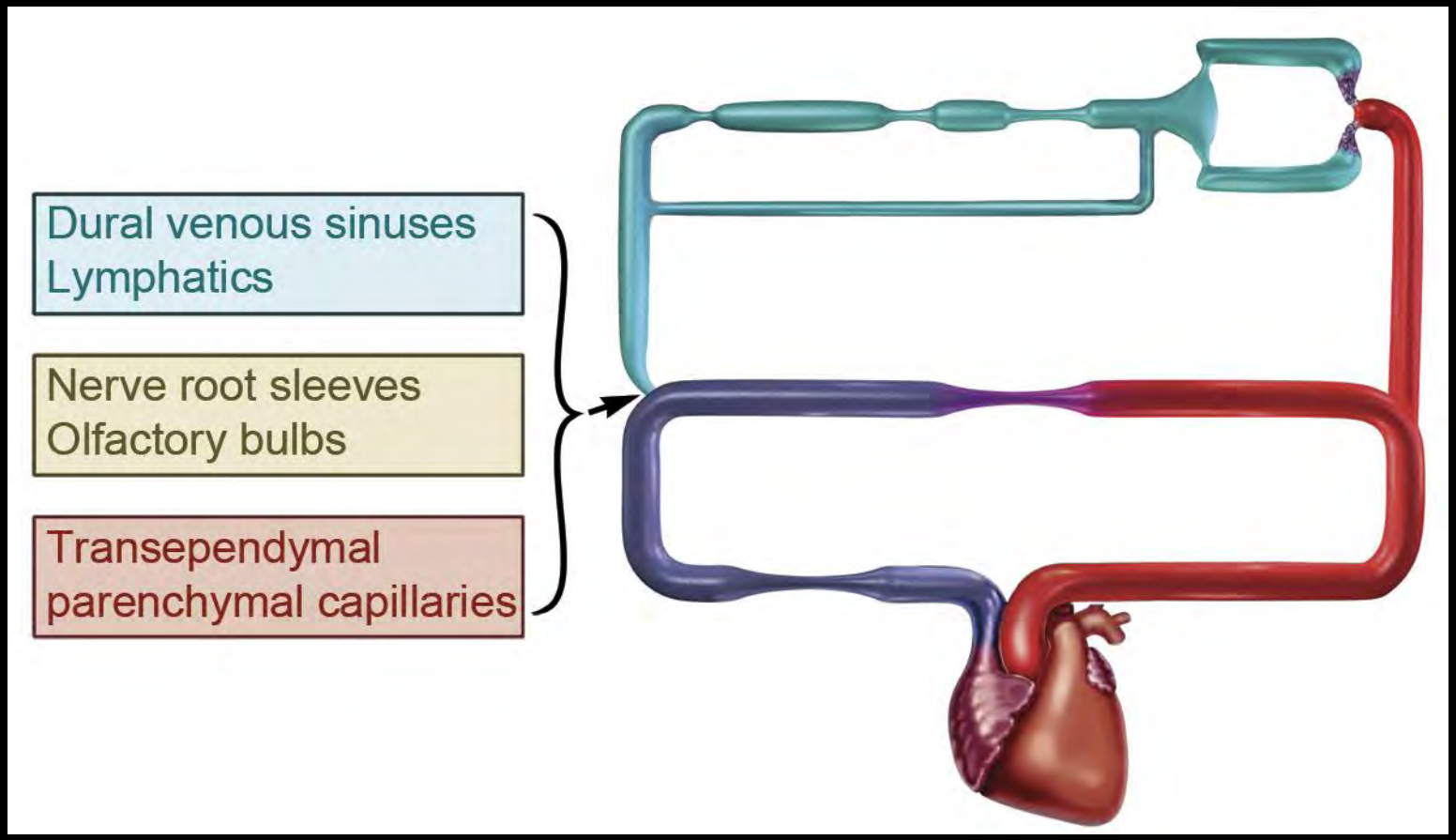
Classification Based On Point Of Obstruction

- 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?



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 Subtantially More Dangerous Than A Shunt Procedure and Subtantially 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