Utility of the new classification of hydrocephalus in the treatment of the "Shunt From Hell"

Where is the point of obstruction?

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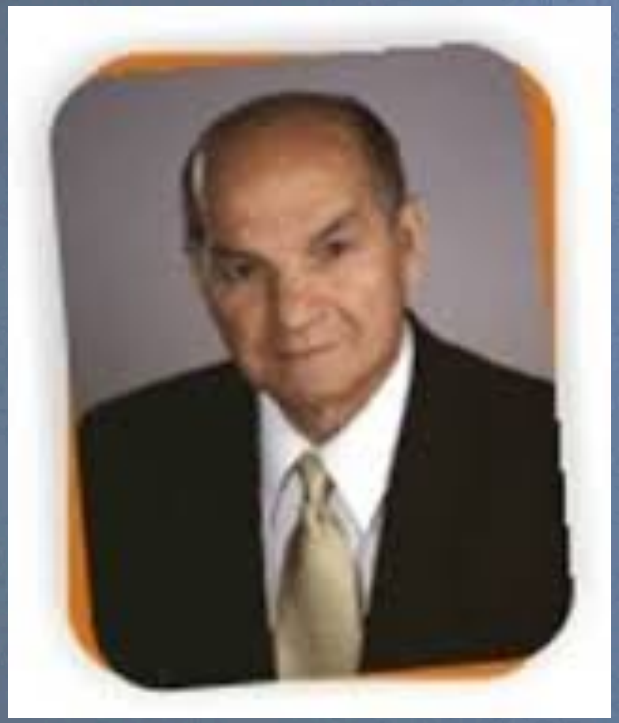
I have no conflicts of interest in regard to this presentation

Conversation with Tony



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"a dynamic imbalance between the formation (production) and absorption of cerebrospinal fluid resulting in ventricular dilatation".

Results of Meetings of neuroscientists in Phoenix and in LA at the ISPN meeting in LA and representing a hard won consensus of essentially pediatric neurosurgical investigators

Discussion:Disorders of CSF

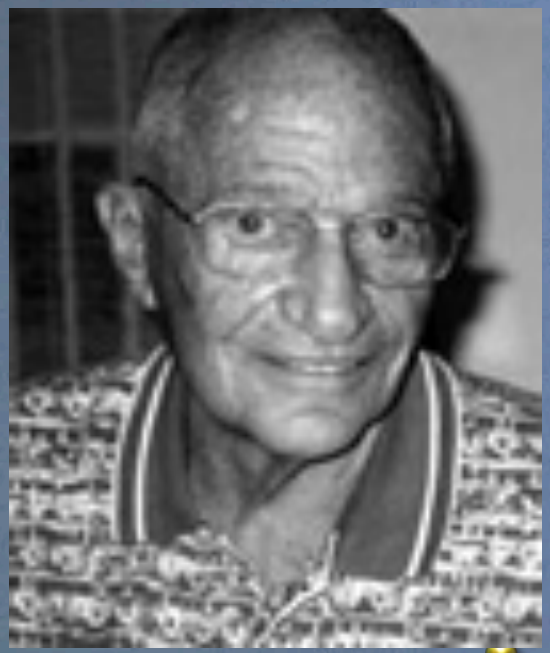
- Included
  - Hydrocephalus
  - NPH
  - INPH
  - ? Normal Volume Hydrocephalus: Began with enlarged ventricles
- Excluded:
  - IIH/Pseudotumor
  - Arachnoid Cysts
  - Cerebral Edema
  - “External” hydrocephalus
  - *Hydrocephalus ex vacuo*

Currently Accepted Classification

Dandy 1913

- "Communicating vs Non-Communicating"
- Tools Available
  - Injection of supravital dyes into ventricle and recovery by lumbar puncture
  - Post-mortem
- Results
  - Choroid plexectomy
  - Third Ventriculostomy

Ransohoff Classification: 1960



- Still only two categories
  - Intraventricular obstructive
  - Extraventricular obstructive
- Tools Available
  - Supravital dyes
  - Air
  - Pantopague
  - RISA
  - Shunts
- Outcome
  - Largely ignored
  - Stimulated experiments
  - Recent work: McAllister

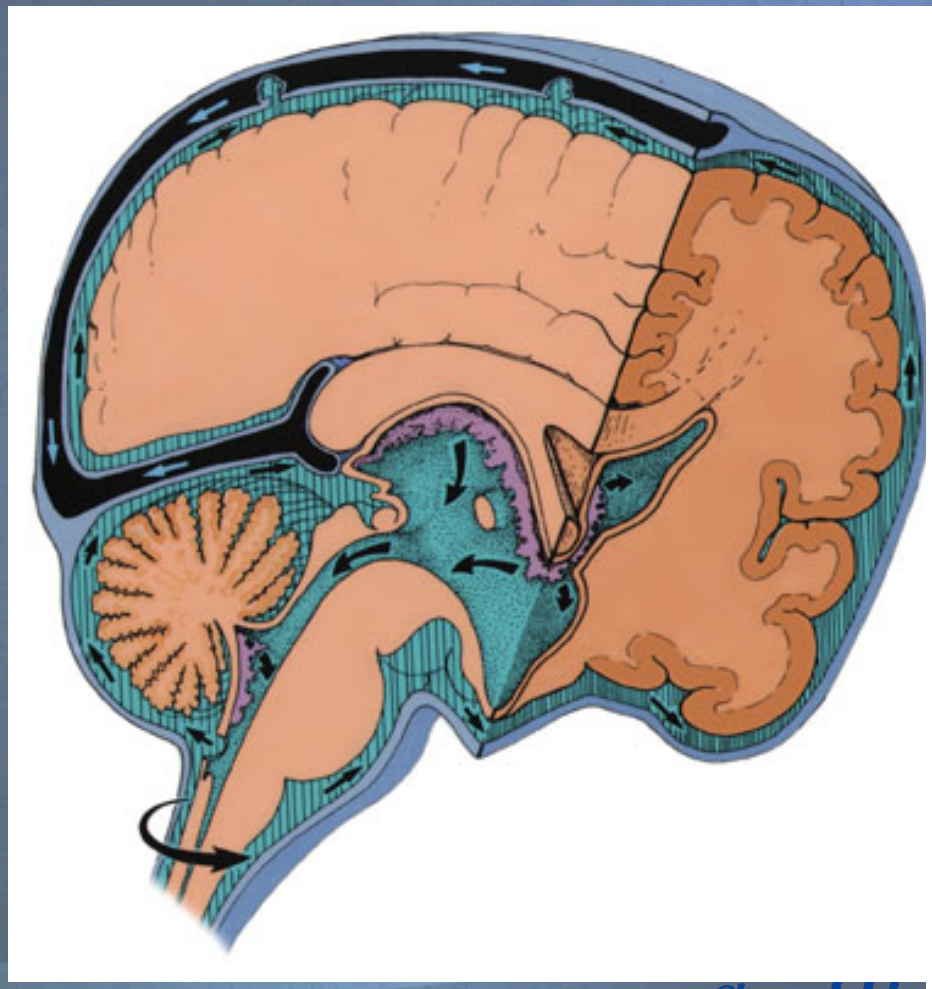
The Consensus Group

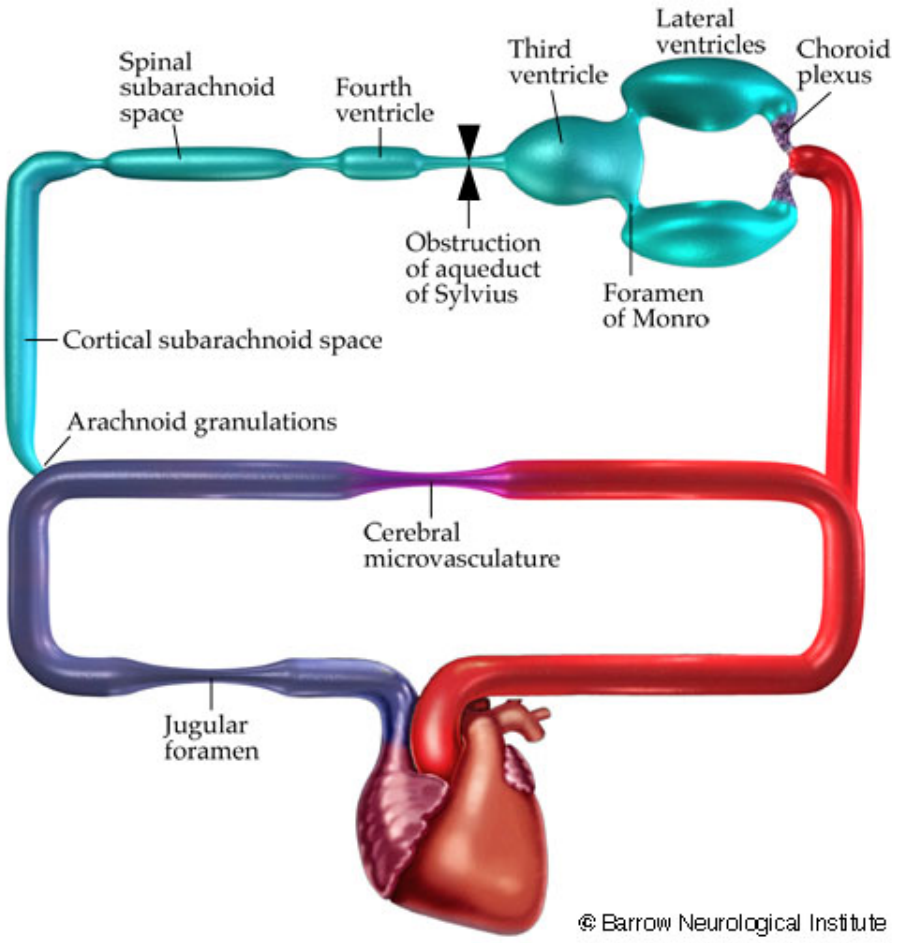
Hajime Arai MD	Tokyo, JP
Concezio Di Rocco	Rome, IT
Petra Klinge	Providence, RI
Conrad Johannson	Providence, RI
Pat McAllister	St Louis, MO
Gordon McComb	Los Angeles, CA
Shizuo Oi	Tokyo, JP
John Pickard	Cambridge, UK
Michael Pollay	Sun City, AZ
Osammu Sato	Tokyo, JP
Spyros Sgouros	Athens, Gr
Marion Walker	Salt Lake City, UT
Harold L. Rekate	Great Neck, NY

Newer Tools Leading to New Classifications

- CT scans
- Cisternography
- MRI
- Cine MRI
- Direct Endoscopic Inspection
- Responses to treatment
- Cooperative Studies
- Evolving MRI techniques to quantify flow

Circuit Diagram





Etiology Based on Point of Obstruction (Classic Obstructive)

- None
- Foramen of Monro
- Aqueduct of Sylvius
- Outlet Foramina of Fourth

- Choroid Plexus Tumor
- Agenesis of F of M
- Tumor (SEGA)
- Cyst,,
- Functional
- Tectal tumor
- Hydrocephalus
- Shunt overdrainage
- Chronic Meningitis
- Tumors

Etiology in Extraventricular Obstructive Hydrocephlaus

Spinal to cortical subarachnoid space

- Late in subarachnoid hemorrage
- Bacterial and viral meningitis

Terminal absorption

- Congenital anomalies

Venous outflow issues

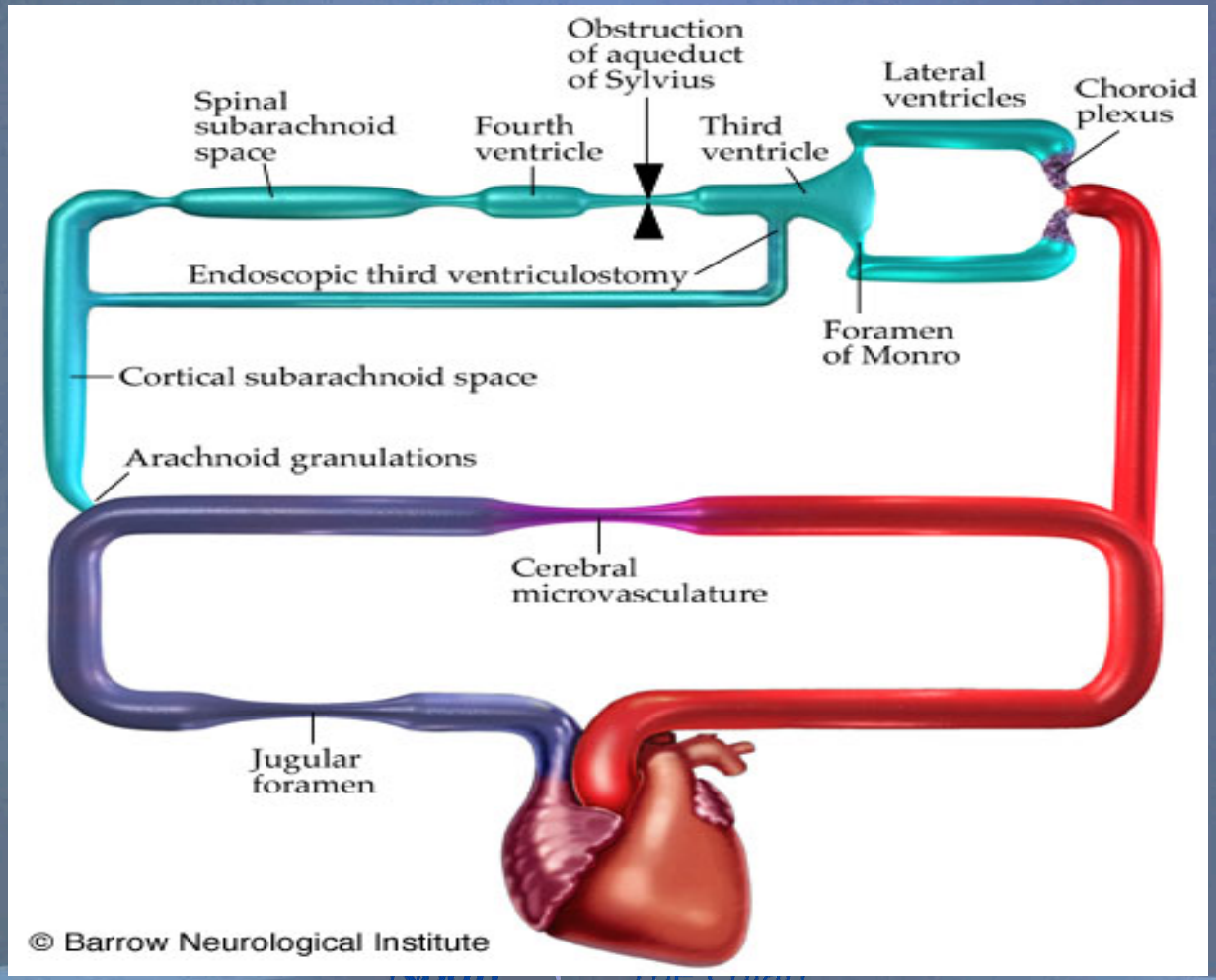
- Early in SAH or infection
- Only causes big ventricles in babies
- Results in pseudotumor or SVS

Treatment Options

- Foramen of Monro
- Aqueduct of Sylvius
- Outlet of fourth ventricle
- SSAS to CSAS
- Terminal absorption
- Venous outflow resistence

- Shunt with 2 catheters
- Septum pellucidotomy
- ETV
- Shunt
- Direct opening of the 4th
- ETV
- VP or LP Shunt
- ETV
- VP or LP
- LPS
- Dural venous stent

What Is Accomplished by Performing an ETV?



The point of obstruction isn't always obvious

- The IHIWG
- Called MRI flow group by the ASNR
- The important work from Dr. Ari Blitz to define the point of obstruction
- Time Slip and other methods for determining the point of obstruction

"A scientific truth does not triumph by convincing its opponents and making them believe but rather because its opponents eventually die and a new generation grows up that is familiar with it."

Max Planck: the father of quantum physics

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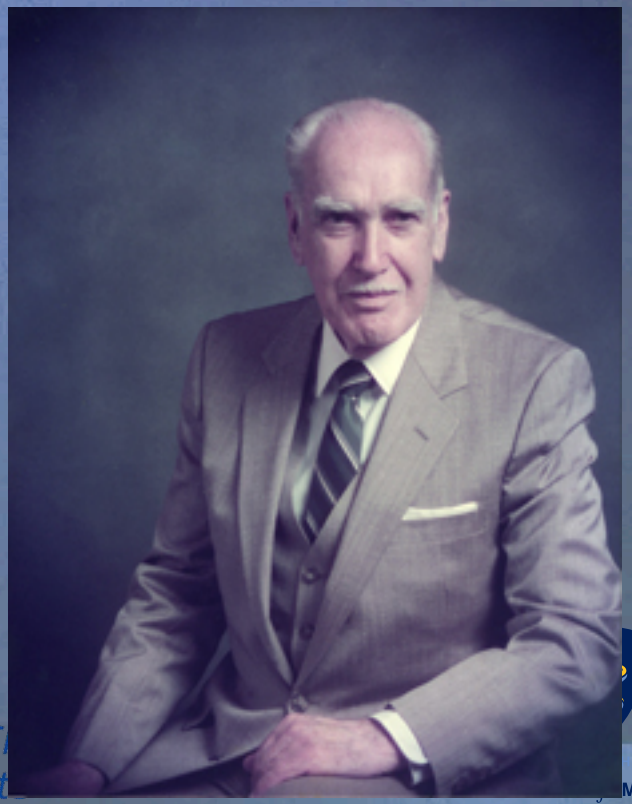
The Essential Role of Neuroradiology

- Recognize and report the likely point of restriction or obstruction of flow
  - Can be challenging in the original diagnosis of hydrocephalus, particularly in babies
    - In infantile hydrocephalus it is common to find multiple points of failure of flow
  - Should be quite simple at the time of shunt failure
- Help develop techniques to accurately identify where flow is impeded
  - Especially in pediatrics there is a distinct reluctance to perform studies involving tracers

Case Presentation
Patient with Dandy Walker Malformation and Hydrocephalus Diagnosed Prenatally



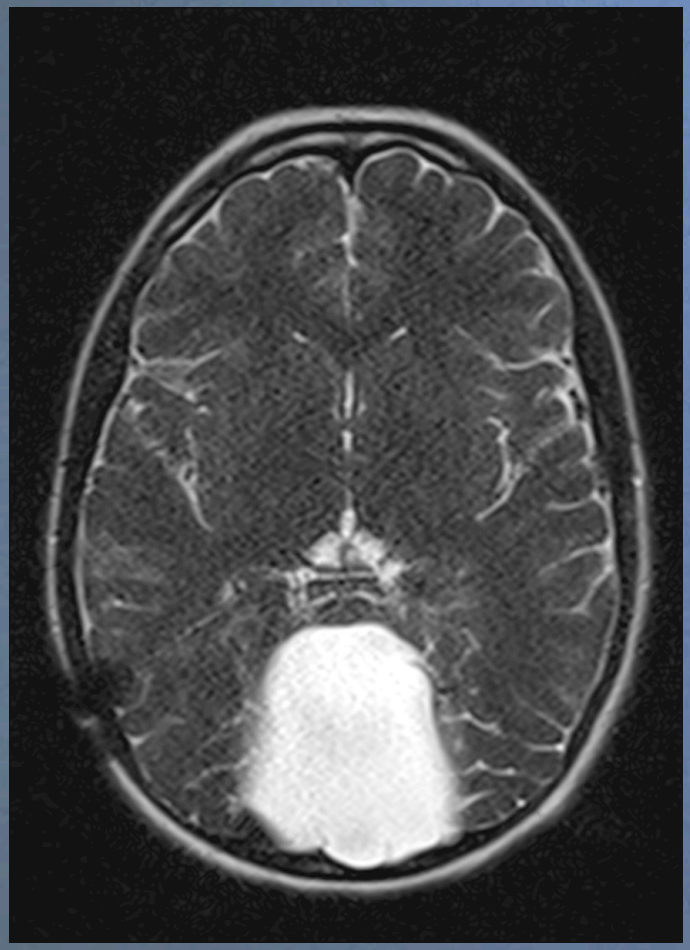
15 year old sophomore shunted in infancy with over 60 shunt revisions

- Diagnosed in utero and family rejected abortion
- First operation was shunt of DW cyst at 2 weeks
- At two years the shunt was converted to a ventriculoperitoneal shunt
- Two shunt failures until age 13
- In the last two years she has spent 5 months in hospital and had 46 shunt revisions

Some things about Amelia

- Neurologically normal including balance
- Honors student when she can actually go to school
- Accomplished guitarist, singer and song-writer (Country and Western)
- Maintains a sense of humor

MRI at crisis period at home



- Large cyst
- CSF in cisterns and cortical subarachnoid spaces
- Large Dandy Walker cyst
- Flow study was read as communication between the third ventricle and the cyst
- I was asked to consult as to what valve to use

CT scan leading to travel to LI

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Rekate's rules for managing the "Shunt from Hell!!!!!!!"

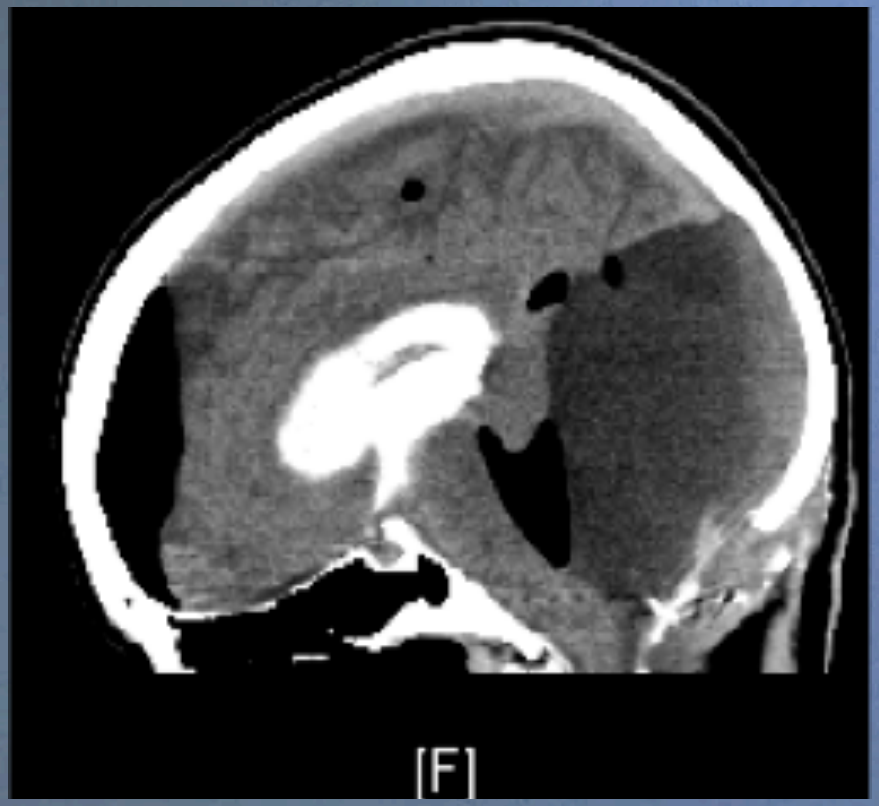
1. All CSF compartments must see the same ICP
2. ICP in supine position 5-15 mmHg
3. ICP in erect position -5-+5 mmHg
4. It isn't a shunt it's a sentence

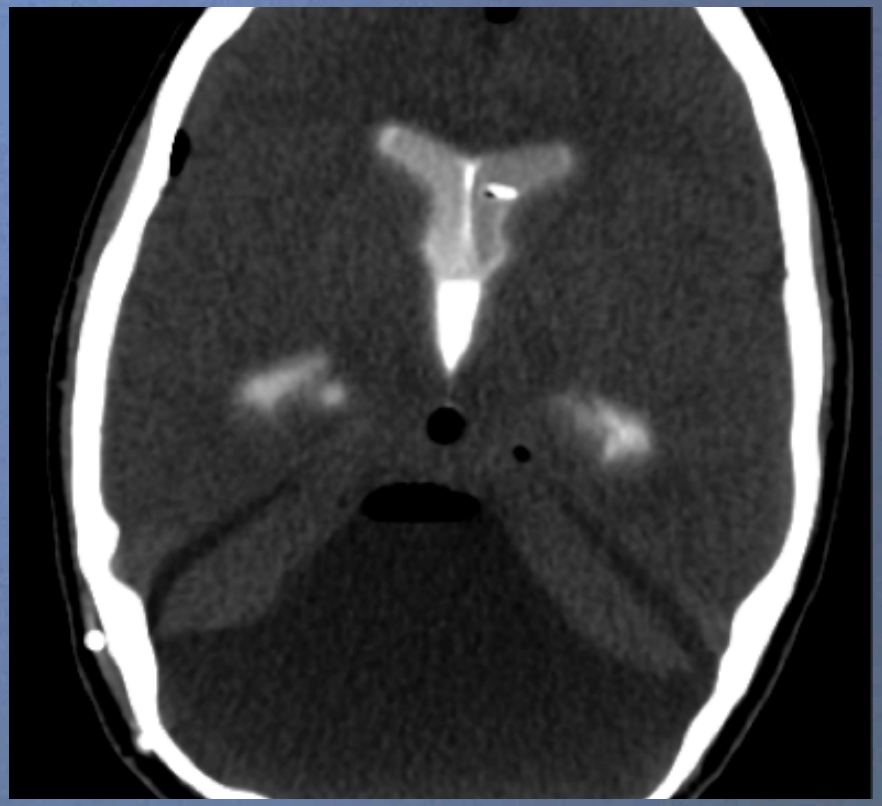
Any Ideas?

Surgical procedure number 1

- Fenestration of DW Cyst
- Cyst to ventricle to peritoneal shunt
- Used Edwards-Barbera "T" shunt to shunt both the cyst and the subarachnoid space

Operation 2 addition of reservoir

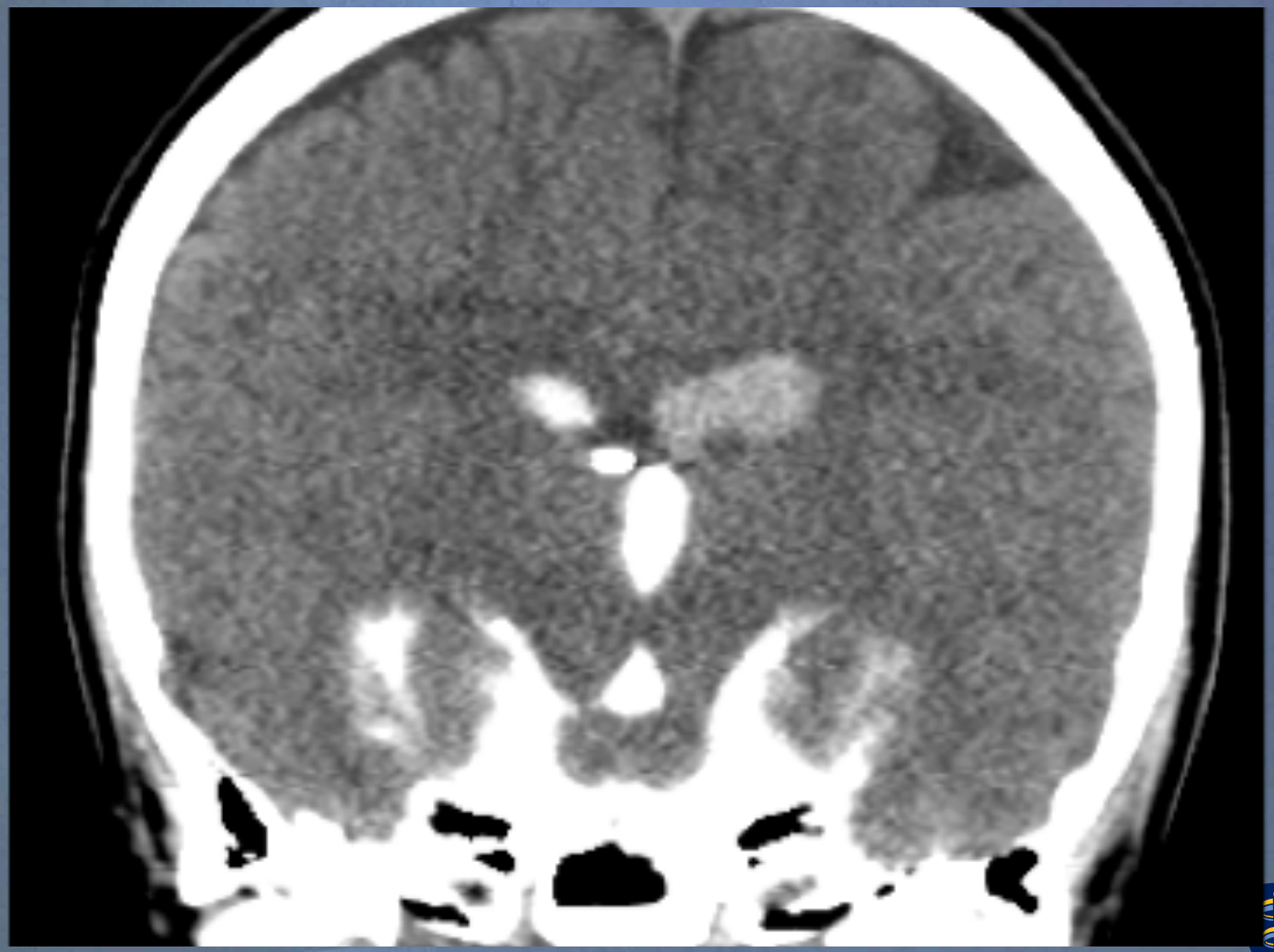




Are We Finished?

- Intracranial pressure monitoring is normal
- Severe headaches persist
- All compartments in communication
  - Communication depends on continued function of three proximal catheters
  - She still has headaches
  - Her assessment of outcome is not great

Operation 3 ETV and tie off shunt



Operation 4: removal of shunt except reservoir