Is The Brain Really Necessary?

What do we know about the importance of the volume of the ventricles, raised intracranial pressure, and mass of the brain

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I have no conflicts of interest in regard to this presentation
I will not be discussing off label use of drugs or devices

Wrapping of the head with an ace bandage as treatment for post-hemorrhagic hydrocephalus-1973

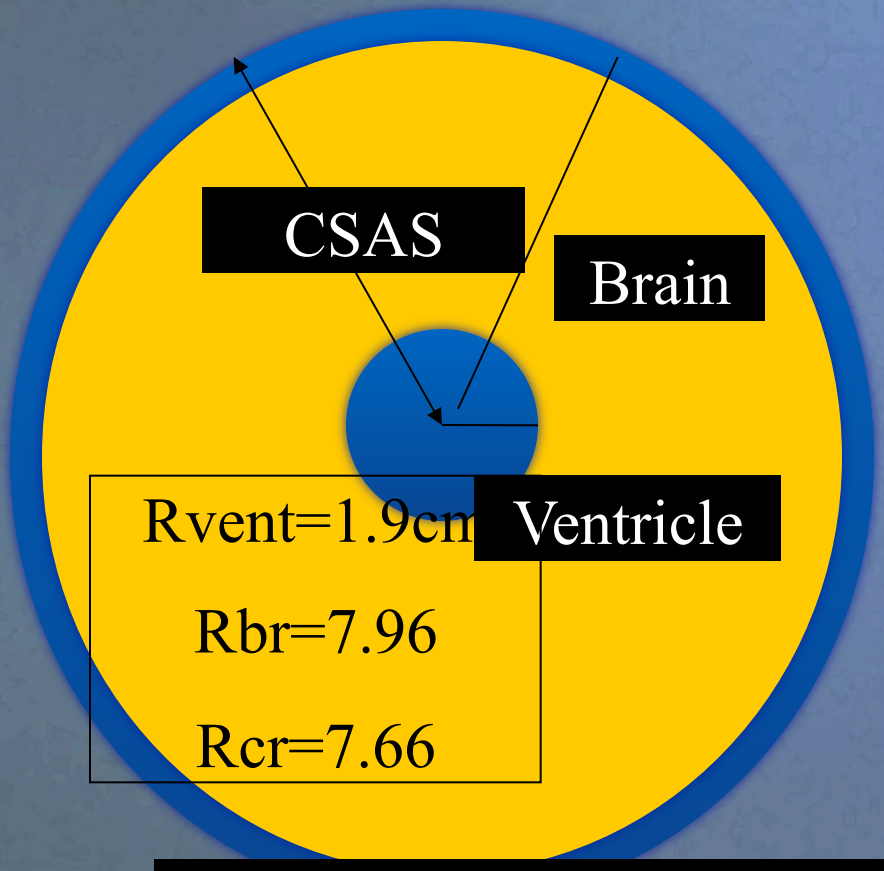
- Hydrocephalus is not an all or none phenomenon
- We have no long term outcome studies on untreated patients known to have hydrocephalus since the early 70s
- What does hydrocephalus do to the brain?
- What causes the neurologic, developmental and cognitive problems we see in these patients?

Lorber observations 1980

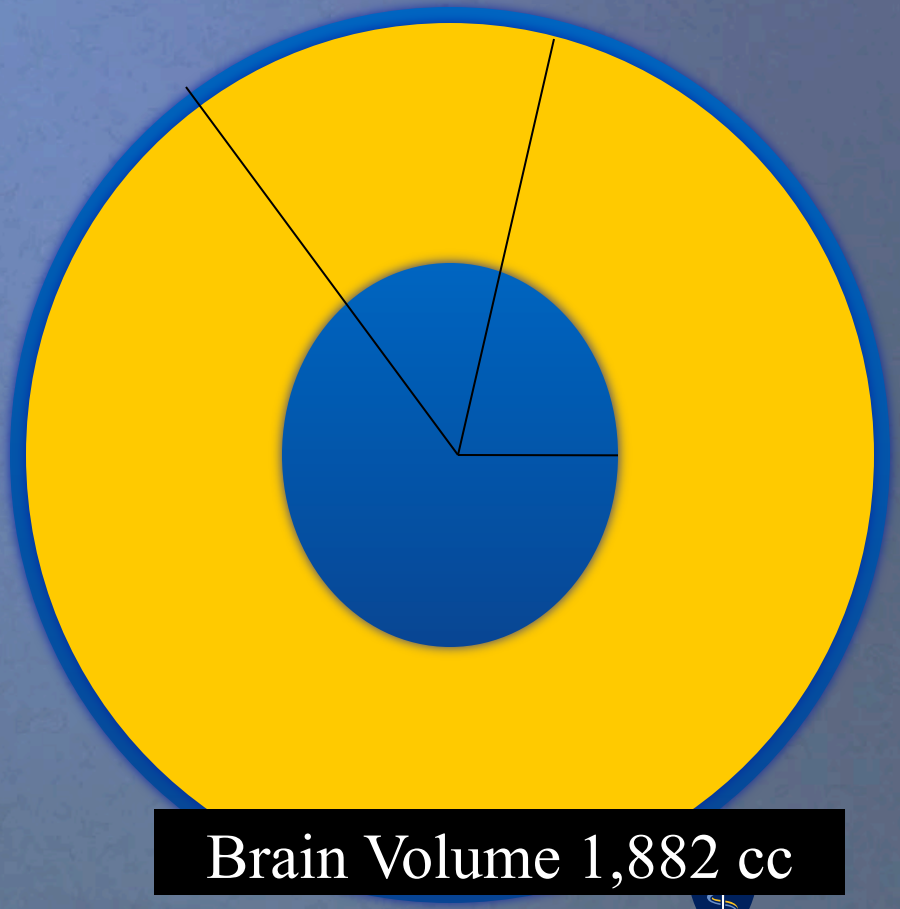
Assumptions And Model

- Volume of normal ventricles-30cc
- Thickness of cortical subarachnoid space 3mm: Intracranial volume-(brain +ventricle)
- What would happen to ventricle volume if the brain is displaced outward to displace cortical subarachnoid space?
- No change in brain volume

Blocked Shunt: Short Time "T"



Brain Volume 1,882 cc



Brain Volume 1,882 cc

Significance Of Ventricular Expansion At Time Of Failure

- There is an obstruction of flow of CSF between the ventricles and cortical subarachnoid space
- The patient is a good candidate for endoscopic third ventriculostomy!!!!!
- The obstruction is not at arachnoid villi
- There is a high likelihood for neurologic recovery

The March To Normal Pressure Hydrocephalus

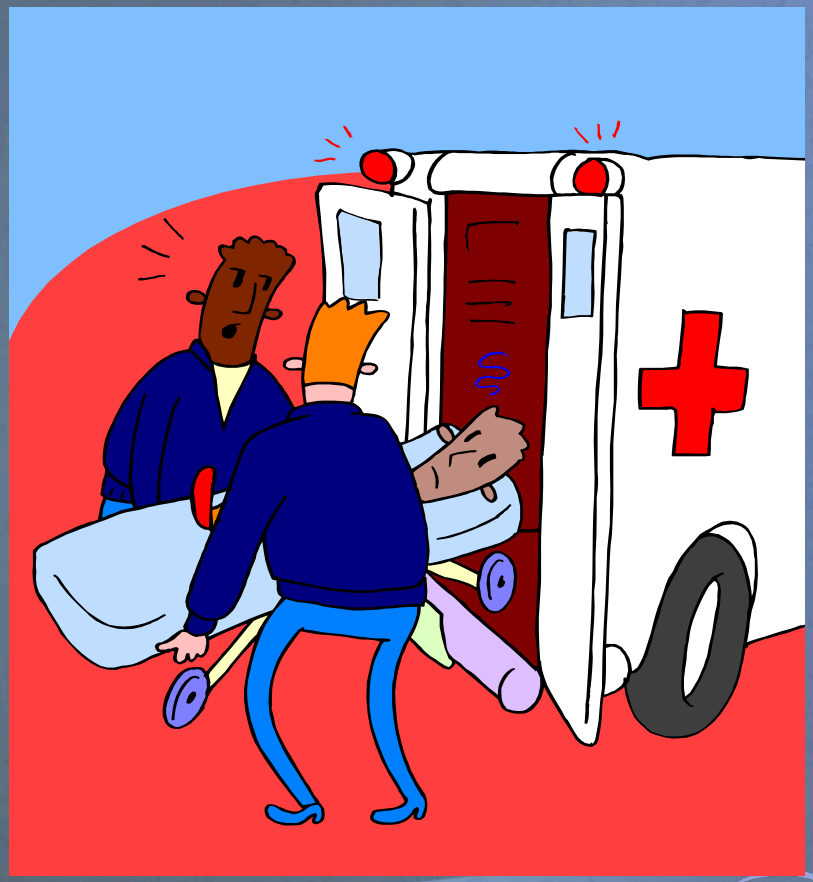
- Chronic Compensated Hydrocephalus
- ***L***ong ***S***tanding ***O***vert ***V***entriculomegaly Of the ***A***dult (***LOVA)***
- ***S***ymptomatic ***H***ydrocephalus ***M***anifesting in ***A***dolescents and ***Y***oung ***A***dults (***SHYMA***)
- ***N***ormal ***P***ressure ***H***ydrocephalus (***NPH***)

Benefits Of Shunting

- Decreases intracranial pressure, decreasing headaches
- Prevents herniation
- Improves cognitive function*
- Prevents subtle deterioration*

*Possibly

Making The Decision

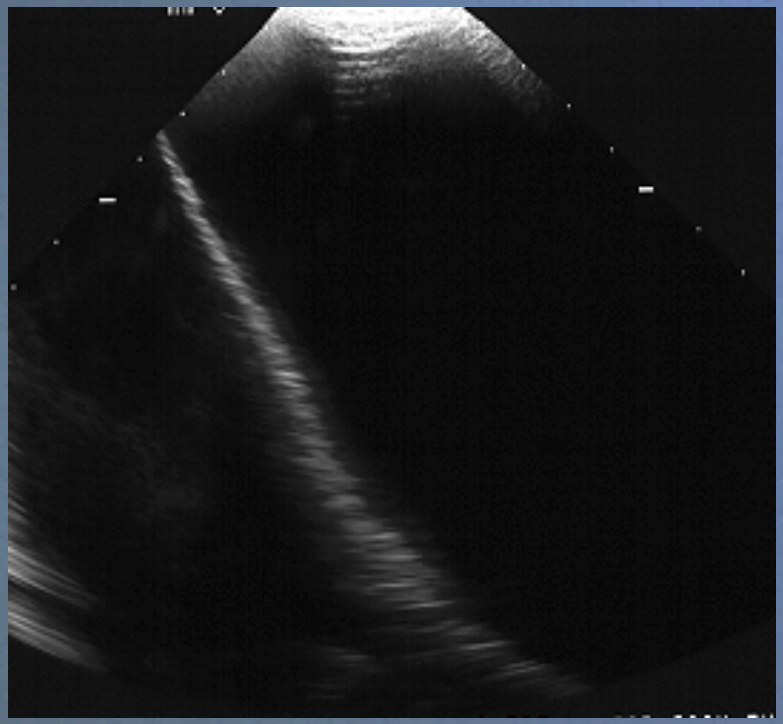


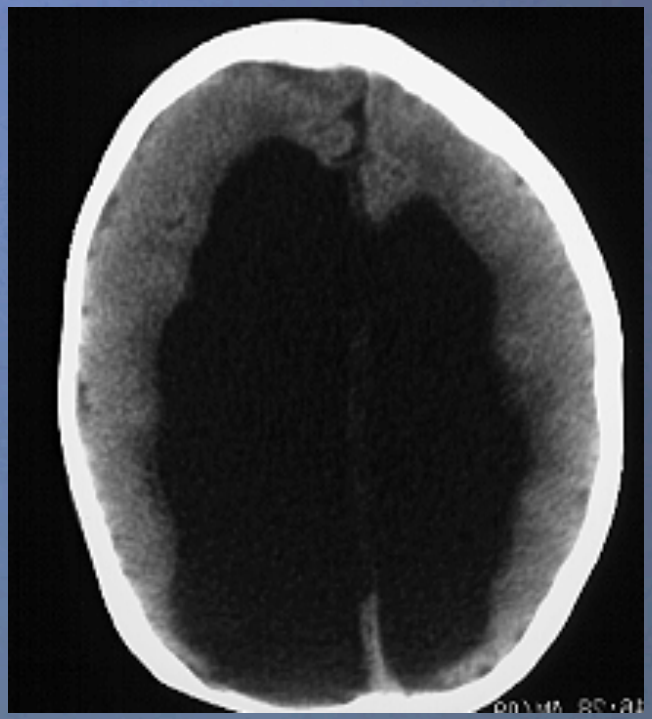
- Unless there is a disorder of level of consciousness there is plenty of time
- Subtle deterioration should be documented
- Incidentally found hydrocephalus is one of our greatest challenges

WHY NOT SHUNT?

- Infection 0-55% depending on population and institution
- Mechanical failure - 40% failure in first year, 60% failure at two years
- Creation of shunt dependency
- Death rate in neonatal hydrocephalus approaches 1%/year

IS THE BRAIN REALLY NECESSARY?





CONCLUSIONS:
note: more than a little prejudice

- In the absence of potential for herniation, overtly increased intracranial pressure or rapid ventricular distention, there is time to ponder intervention
- Deterioration must be documented
- Neuropsychology is important in making decision

CONCLUSIONS II

- There will never be absolute certainty regarding the "black and white" threshold
- Arguments are somewhat similar to those for Caesarian Section
  - Reimbursement for intervention
  - Legal "second-guessing" for non-intervention
  - Almost never criticized for intervention

TWO SIDES OF THE ARGUMENT

- "WHEN IN DOUBT-SHUNT!!!!
- PRIMUM NON NOCERE

Definitions

- Chronic Compensated Hydrocephalus
- Stable moderate ventriculomegaly
- Late deterioration is inevitable
- Patient is being subtly harmed
- Substrate in NPH

- Arrested Hydrocephalus
- Deterioration is not inevitable
- Patient not being harmed
- May be impossible to determine the difference between these two

ETHICAL OBLIGATIONS

- Informed Consent should include discussion of level of certainty
- Obtain neuropsychological workup in "gray zone" cases
- Careful assessment of what science there is prior to participation in the legal process

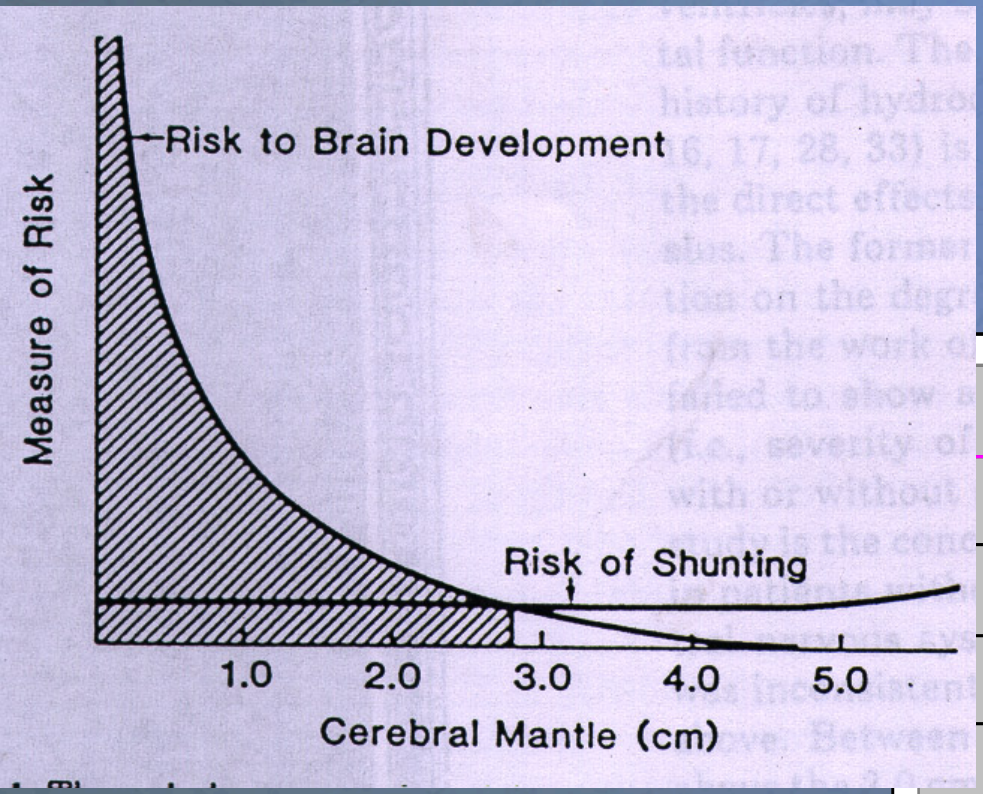
What Is The Cognitive Cost Of Untreated Hydrocephalus

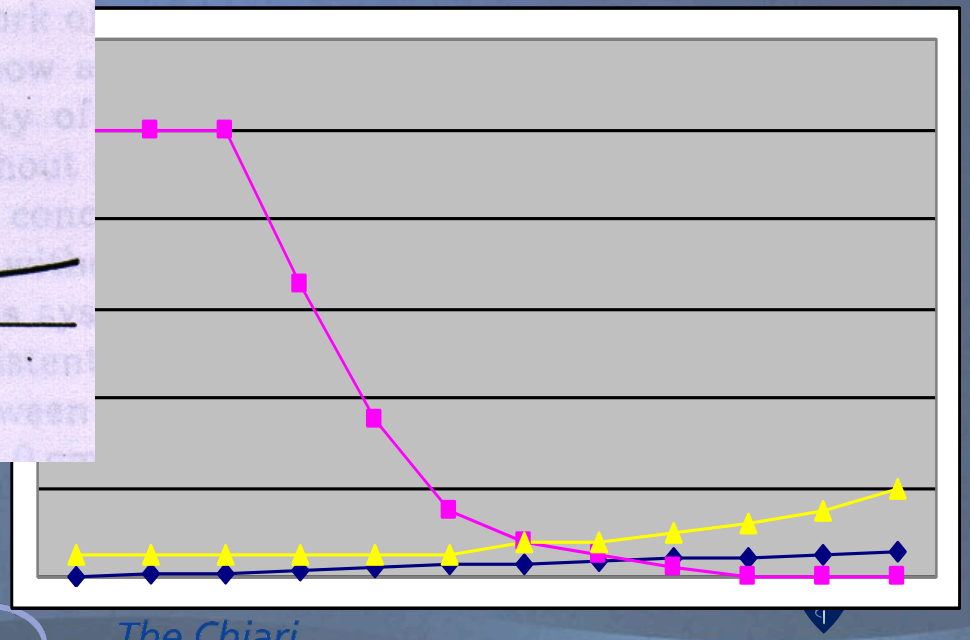
- Many assumptions, Few answers
- Is the cognitive loss in hydrocephalus time dependent?
- Are there any conditions in which hydrocephalus develops without a direct effect on the brain itself?

Hydrocephalus Is Not An "All or None" Phenomenon

- Success in treating hydrocephalus using ETV does not require the ventricles to return to normal.
- Success in treating NPH with shunt does not require return of ventricles to normal.
- Ventriculomegaly in post-hemorrhagic hydrocephalus self-stabilizes or reverses in half of the cases within 4 weeks.

Risk VS Ventricular Size





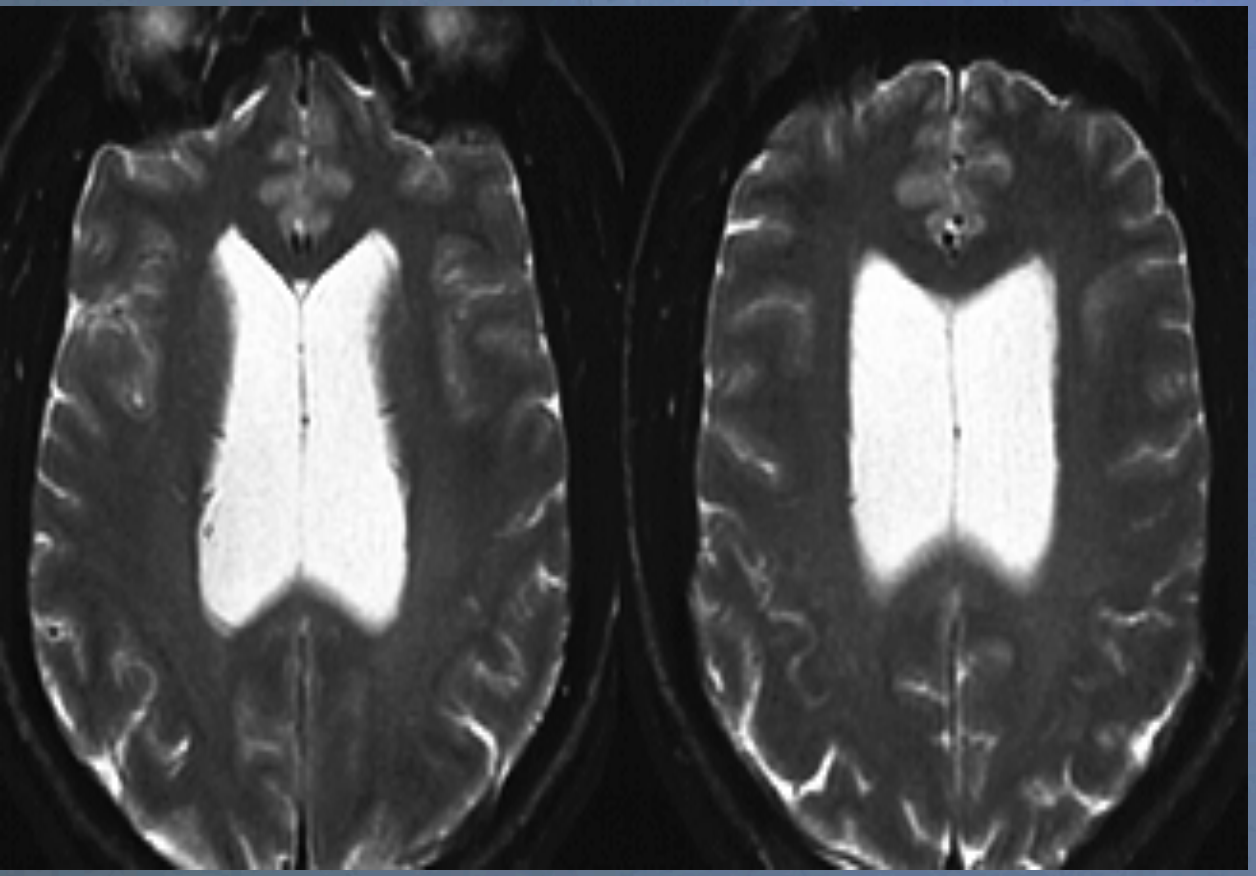
Stable ModerateVentriculomegaly: Chronic Compensated Hydrocephalus

- The Biggest Challenge
- The Fewest Answers
- How Does One Predict the Outcome With and Without Intervention
- The important role of neuropsychology

What We Don't Know About SMV

- How Many IQ Points Are Lost?
- At What Point if any is It futile to treat with shunt or third ventriculostomy?
- Are the risks of death or intellectual deterioration eliminated if the patient is not shunted?
- How does this relate to success or failure of third ventriculostomy?

HOW DO YOU DECIDE?



53 Year old mechanic

Single seizure

Memory difficulties

- He and wife disagree about how severe

No Headache

No Urgency

No Gait disturbances

NEUROPSYCHOLOGY

- Verbal IQ-96 Performance IQ-117
- Normal Attention Span
- Normal dexterity
- Memory normal for the patient's age, educational level (12 years), and IQ
- Problem Solving mean as expected for job and education.
- NO SHUNT PERFORMED/ REPEAT

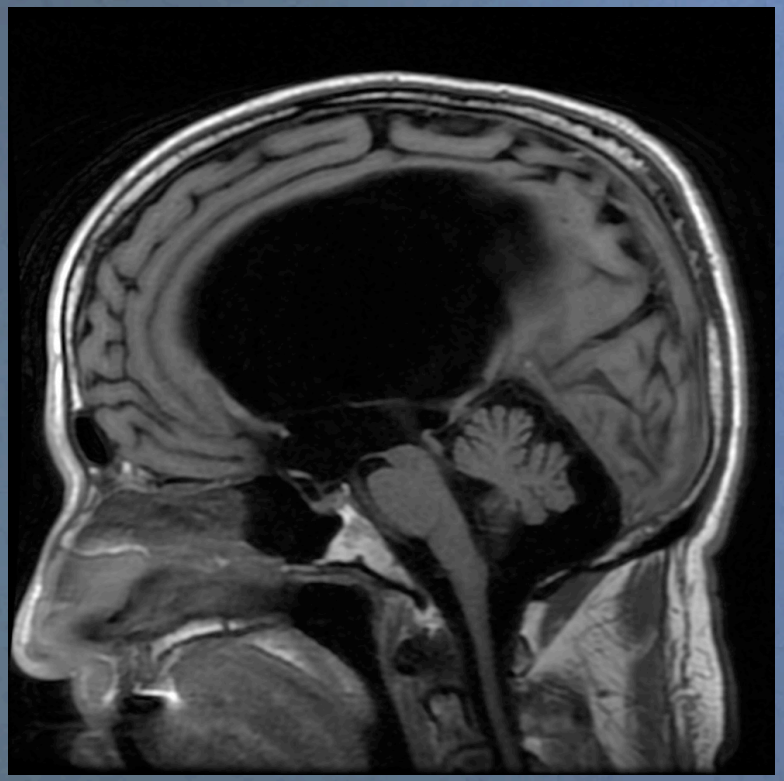
LOVA

- 32 yo Bank Executive
- Brief loss of consciousness following auto accident
- No headache or signs of NPH

Neuropsychology

- Excellent undergraduate degree and performance
- M. BA. At night
- Long history of overachieving
- Palm pilot, legal pad, excellent secretary
- Clear evidence of decrease in performance relative to previous academic performance and genetics
- VP Shunt performed

Have we waited too long?



- 68 year old retired postal worker
- Head circumfirence 65cm
- Father of 3
- Typical NPH triad progressive over the past three years

ETV in LOVA patients

- All aqueductal stenosis isn't
- Headaches are rarely if ever related to LOVA
- 4/6 ended up with shunts

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Longstanding overt ventriculomegaly in adults: pitfalls in treatment with endoscopic third ventriculostomy

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Object. The recently described condition of longstanding overt ventriculomegaly in adults (LOVA) has not been defined in terms of the need for intervention, timing of intervention, and ideal treatment. The purpose of this review was to evaluate the role of endoscopic third ventriculostomy (ETV) in the treatment of LOVA.

Methods. Data collected in six patients with LOVA who had undergone ETV were reviewed retrospectively in terms of the definition of treatment success, rates of success, complications, and outcome. All six patients presented with headache disorders. In all patients, triventricular hydrocephalus had been diagnosed as aqueductal stenosis, and head circumference measurements were above the 98th percentile. All six had undergone successful ETV as documented by the free flow of cerebrospinal fluid into the basal cisterns, which remained open throughout the follow-up period. After the procedure, one patient experienced a mild degree of difficulty with short-term memory. Five patients remained symptomatic or had symptoms requiring further treatment 3 months to 3 years after ETV. Four patients received ventriculoperitoneal shunts, and one underwent venous stenting for high intracranial pressure after successful ETV. In two patients in whom aqueductal stenosis had been diagnosed, the sylvian aqueduct was patent after the procedure.

Conclusions. In LOVA patients who present with headaches, ETV may not lead to improvement in the headaches. Despite the presence of triventricular hydrocephalus, closure of the aqueduct may be a secondary phenomenon, and flow through the aqueduct may be reestablished after ETV. If intracranial hypertension persists after successful ETV, its cause may be increased venous sinus pressure.

KEY WORDS • **aqueductal stenosis** • **compensated hydrocephalus** • **normal-pressure hydrocephalus** • **endoscopic third ventriculostomy**

SHYMA Context

- Large head or evidence of chronic hydrocephalus never shunted
- Patients shunted in childhood with evidence of increase in ventricular size from earlier studies.
- Long asymptomatic period
- Deterioration with incident
  - Minor head injury
  - Medical illness
  - Others

Thank You

North Shore LIJ The Chiari Institute

North Shore-Long Island Jewish Health System

HOFSTRA NORTH SHORE-LIJ SCHOOL of MEDICINE

Negative Pressure Hydrocephalus

- ICP recorded is below atmospheric pressure
- Shunt failure diagnosed but at exploration found to be working
- Deranged consciousness with negative intracranial pressure and enlarged ventricles
- Air is sucked in if the system is opened

Too Little Fluid In CSAS

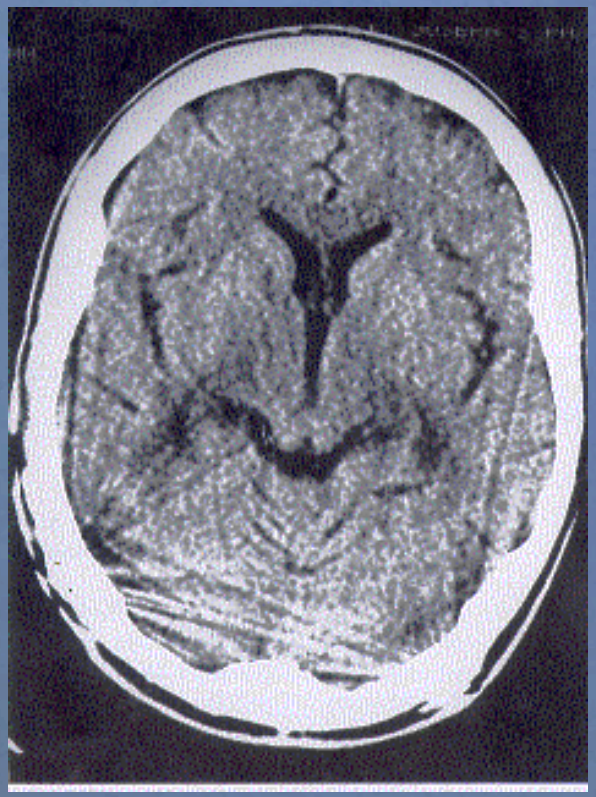
- Spontaneous intracranial hypotension
- Following non-valved lumboperitoneal shunts or syrinx shunts in communicating syringomyelia
- Low or negative pressure hydrocephalus

Negative Pressure Hydrocephalus: Context

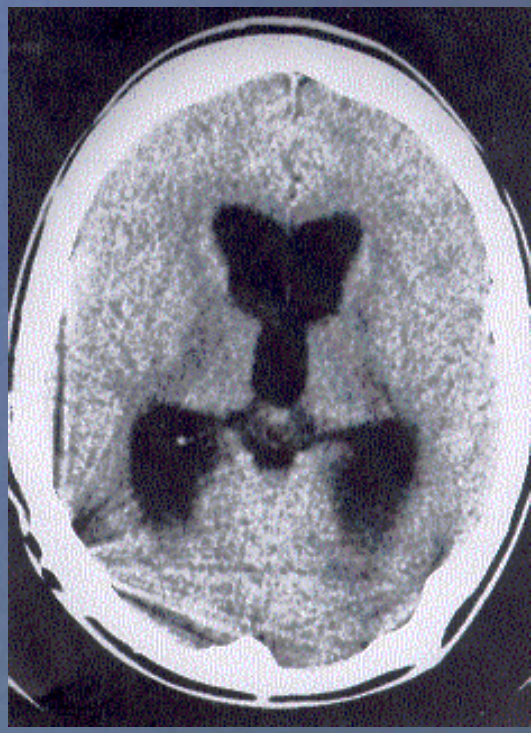
- I have not seen any patients with this syndrome and Know of no patients with this whose hydrocephalus presented in infancy
- All of the patients I have seen (? 25) have developed hydrocephalus in adolescence or adulthood
- All patients have been found to have lack of communication between the ventricle and cortical subarachnoid spaces

Floppy Brain Syndrome

Coma, Low ICP



Cervical Ace Bandage



Ace Removed

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HOFSTRA NORTH SHORE-LIJ SCHOOL of MEDICINE

Context of Low and Negative Pressure Hydrocephalus

- Following skull base surgery either with pre-existing hydrocephalus or complicated by infection
- Patient with hydrocephalus and spontaneous rupture of Tarlov cysts
- Lumbar shunts
- Negative-pressure and low-pressure hydrocephalus: the role of cerebrospinal fluid leaks resulting from surgical approaches to the cranial base.
- Filippidis AS, Kalani MY, Nakaji P, Rekate HL.
- J Neurosurg. 2011 Nov;115(5):1031-7. Epub 2011 Jul 29.
- PMID: 21800965 [PubMed - indexed for MEDLINE]