

Reversible Dementia & The Neurosurgeon

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Dementia

DSM- IV Dementia (abbreviated):

- A. The development of multiple cognitive deficits manifested by both
 1. Memory impairment
 2. One (or more) of the following cognitive disturbances
 - (a) Aphasia
 - (b) Apraxia
 - (c) Agnosia
 - (d) Disturbance in executive functioning

- B. The cognitive deficits in Criteria A1 and A2 each cause significant impairment in social or occupational functioning and represents a significant decline from a previous level of functioning.

Alzheimer's Disease International

Estimates 44 Million have dementia
In 2050 → 135 Million

G8 Summit 2013

Dementia could bankrupt the world's health care systems
with a global cost approaching 1 trillion dollars by 2050 –
Silver Tsunami ??

The Mauritius Alzheimer's Association

- 30 May 2005
- Is a member of ADI (Alzheimer Disease International)
- 2007 → 6000 persons had Alzheimer disease

Statistics Mauritius

- 2052
30.2% of the pop. > 60 yrs old
- 2012
Fertility Rate : 1.41
Recommended : 2.1
- Life Expectancy : 73.64 yrs
- 13.2 newborn per 1000 in 2007
11 newborn per 1000 population in 2012

FAST AGEING POPULATION ??

Common causes of reversible dementia

- **Brain disease**
 - Tumors
 - Subdural hematoma
 - Hydrocephalus
- **Depression**
 - Response to life's stresses
 - Chemical imbalances in the brain
- **Medication**
 - Negative drug interactions
 - Drug overdose
 - Alcohol abuse
- **Malnutrition**
 - Vitamin (A, C, B-12 and folate) deficiencies
 - Mineral (iron) deficiencies
- **Heart disease -- Lack of oxygen to the brain causes confusion**
 - Arrhythmias
 - Congestive heart failure
 - Myocardial infarction

- **Traumas**
 - Usually due to falls
 - Concussions (skull fractures) or contusions (bruises) to the head

- **Metabolic or endocrine disorders**
 - Thyroid disease
 - Hypo/hyperglycemia and other electrolyte imbalances
 - Dehydration
 - Accidental hypothermia
 - Renal failure
 - COPD (Chronic Obstructive Pulmonary Disease)

- **Infection**

Produces fever, affecting brain's cognitive abilities

- **Environmental changes**
 - Visual and hearing loss
 - Loss of daylight and decrease in activities can result in "sundowning"
 - Heavy metal poisoning from gas leaks, exhaust fumes or other toxins

Common causes of irreversible dementia

- Result in permanent Brain Damage
- Cannot be reversed or cured

- **Alzheimer's disease**

- Fourth Leading cause of death in United States

- **Vascular dementia or multi-infarct Dementia**

- **Other neurological diseases causing irreversible dementia**

- Parkinson's Disease
- Huntington's Chorea
- Pick's Disease
- Creutzfeldt-Jacob Disease
- Down's Syndrome
- AIDS

***Is there such thing
as reversible or
irreversible
Dementia?***

Normal Pressure Hydrocephalus

The Reversible
Dementia!!

Adams and Hakim (1965)

Normal Pressure Hydrocephalus (NPH)

- Progressive **dementia**
- Unsteady **gait**
- Urinary **incontinence**
- CSF Pressure below 200 mm

NPH Clinical Diagnostic Criteria

Major criteria:

1. **Ventriculomegaly**
2. **Movement disorder:** with one or more of the following features:
 - a. magnetic / shuffling gait
 - b. abnormal standing base
 - b. bradykinesia
 - c. limb apraxia
 - d. retropulsion
 - e. hypometric turning
 - f. forward leaning posture
 - g. decreased manual dexterity

3. Incontinence: with at least one of the following features:

- a. Urgency
- b. Frequency
- c. Indifference

4. Cognitive Impairment: with one or more of the following features:

- a. attentional impairment
- b. diminished insight
- c. impaired learning and recall
- d. behavioral alterations

Incidence/Prevalence

- 6% of all cases of Dementia
- Studies (Nursing homes) : 9 to 14%

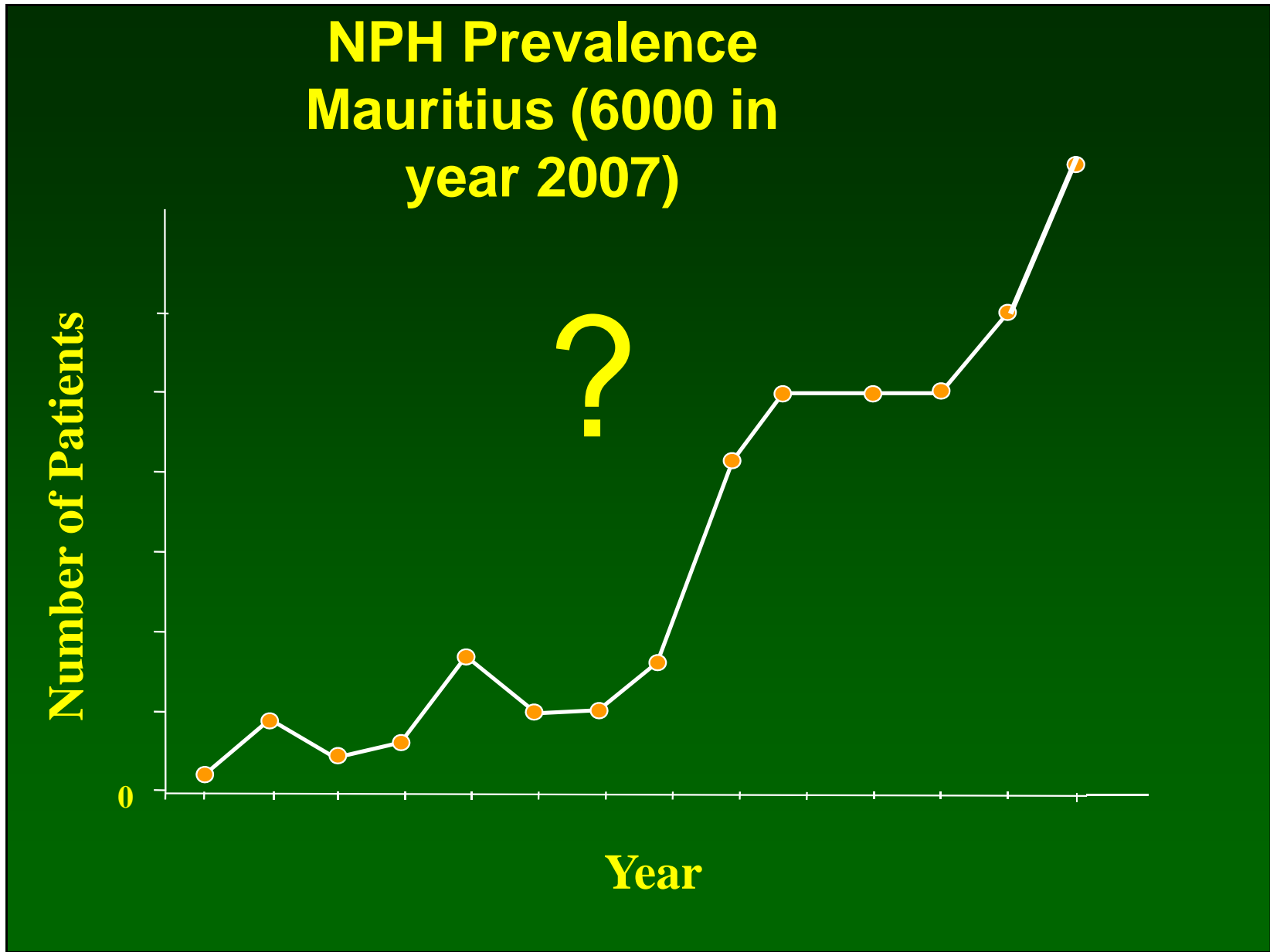
- At Anywhere Incidence = 0.2 to 5.5 new cases per 100, 000 persons per year

- Prevalence – 0.003% under 65 yrs
 - 0.2% to 2.9% > 65 yrs

Study of NPH Risk Factors in Nursing Home Patients

N = 180

Incontinence	75.3%
Non-ambulatory	86.0%
Dementia	45.9%
Depression	46.5%



Pathophysiology (multiple theories)

- Ischemia of deep white matter
- Increased resistant to CSF absorption
- Alteration at the site of absorption
- Hyper dynamic aqueduct CSF flow
- Reduced compliance of subarachnoid space
- Overall reduction of cerebral blood flow

Radiological Diagnostic Criteria



Thinning of Corpus Callosum



CSF Flow Void in Aqueduct



**Periventricular Hyper intensity (CSF Trans-Ependymal
Sepage)**

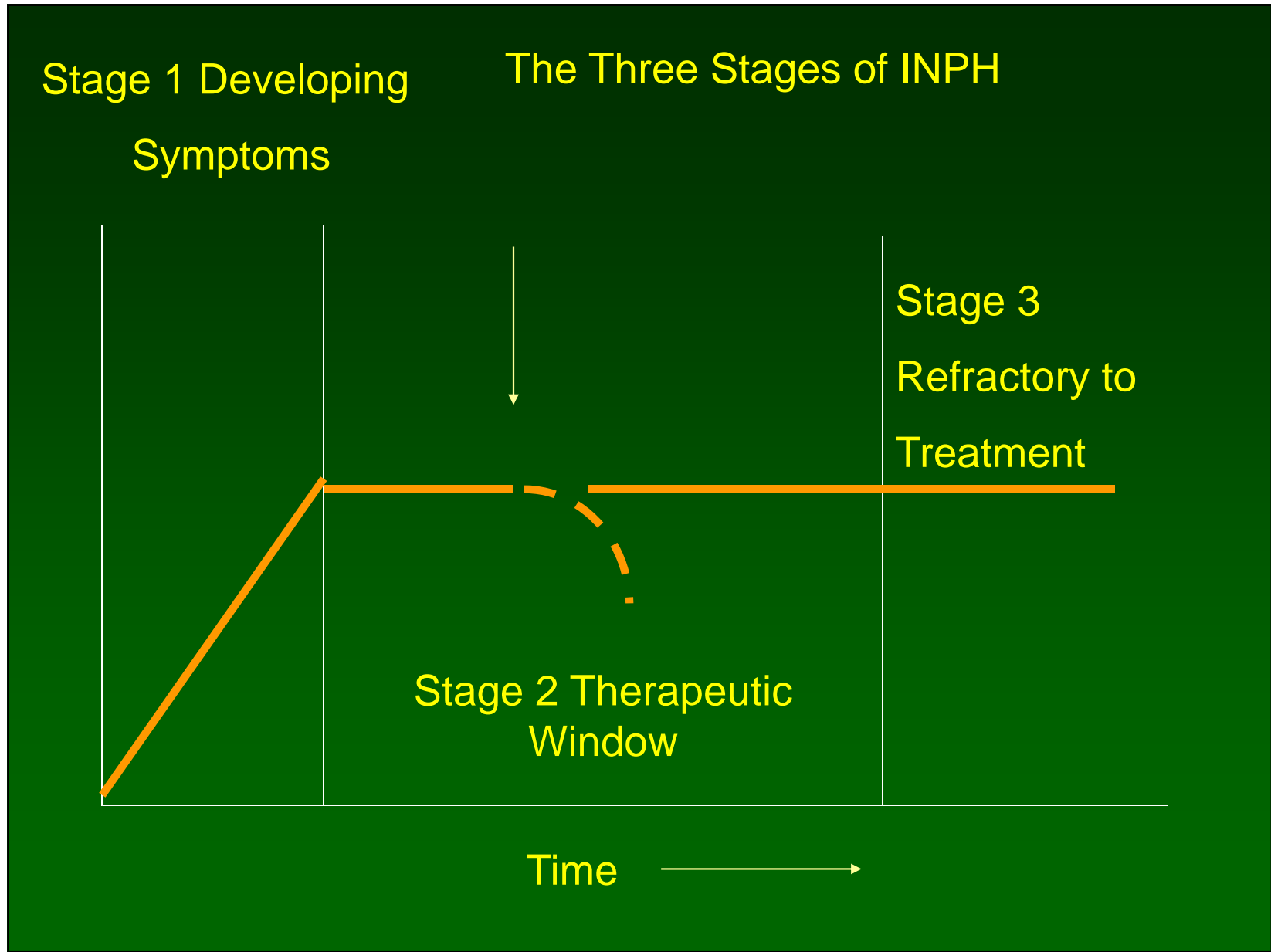


Differential Diagnosis

- Senile Dementia
- Alzheimer's Disease
- Parkinson's Disease
- Multi infarct Dementia
- Depression
- Spinocerebellar degeneration
- Spinocerebellar degeneration due to alcohol

Misdiagnosis/Under Diagnosis

- Less than 350 shunt insertion for NPH in UK – which represents 10% of expected incidence!!
- Failing to entertain NPH as a diagnosis even when faced with a typical patient or looking for the typical patient before entertaining NPH as a diagnosis
- NPH can exist in patients who have only some of the clinical or radiological finding
- Thorough evaluation is required
- Don't deny option of shunting prematurely



Diagnosis of NPH

The diagnosis of idiopathic normal-pressure hydrocephalus remains controversial, particularly in selecting patients for shunt insertion.

Diagnostic Tests

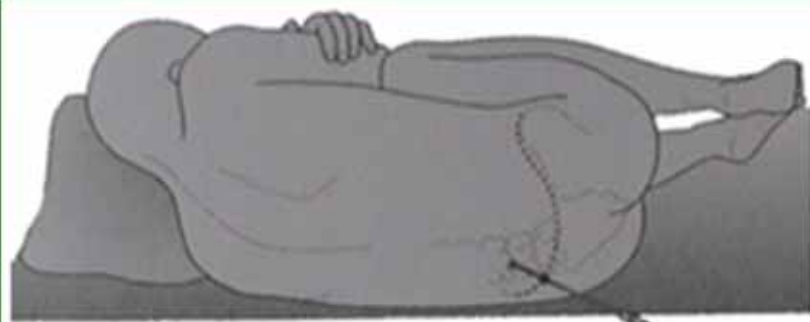
- **ELD (External Lumbar Drainage)**
- **Lumbar Tap (TAP 30/30)**

1. External Lumbar Drainage (ELD)

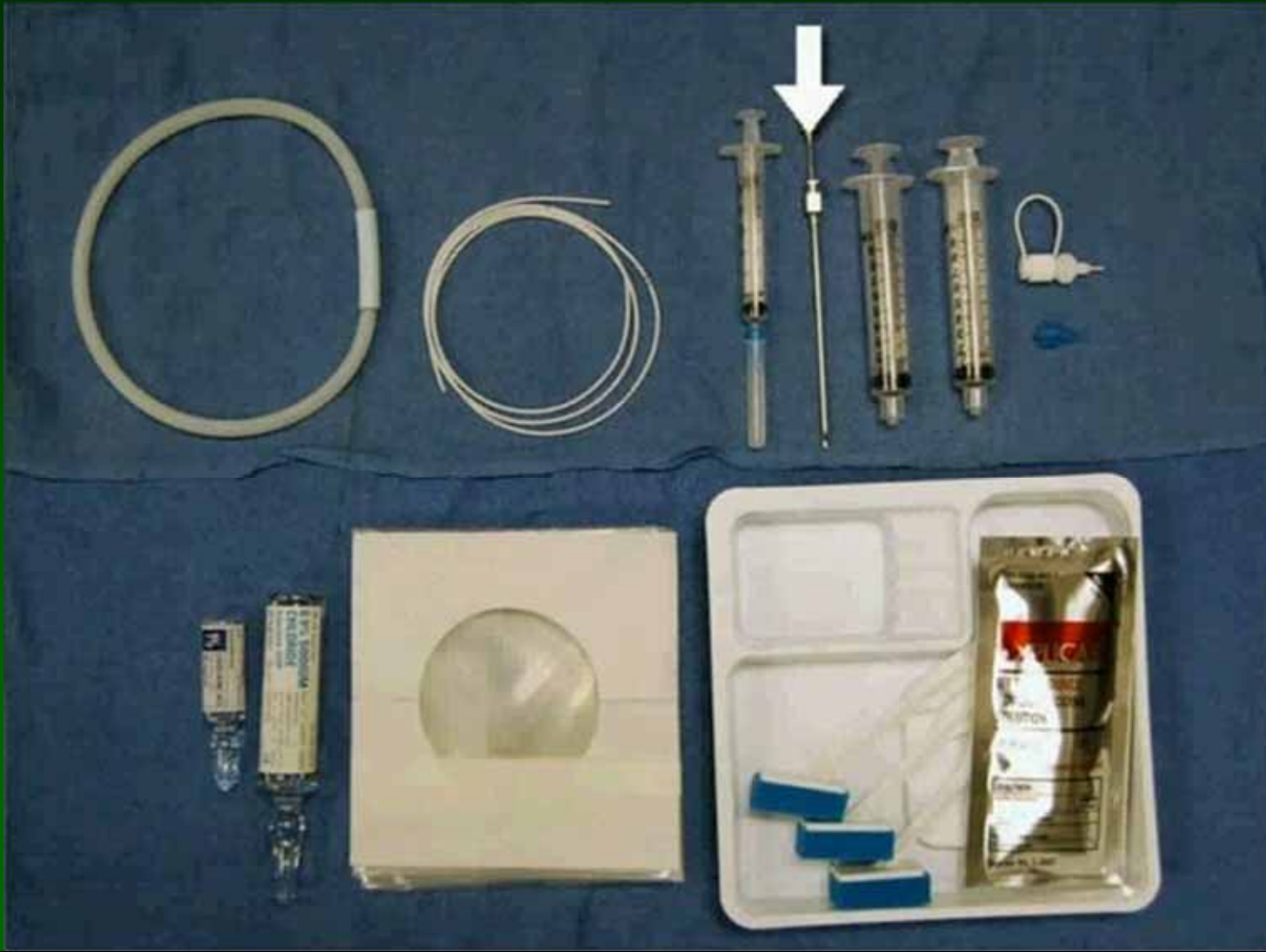
Primary: Prolonged 3 day drainage of patients with suspected NPH CSF will be of prognostic value

ELD (External Lumbar Drainage)

External lumbar drain inserted and CSF drained over a period of three days and patient is assessed clinically



External Lumbar Drain Set



Protocol for NPH Patient

Clinical Evaluation (Outpatient)

(1) Symptom; Gait Disturbance, Memory Loss, Urinary Incontinence

(2) Radiological Finding; Head CT, MRI, Spinal MRI, Spectroscopy

Clinical Diagnosis of NPH

Three Days Drainage (Admission)

Improve after Drainage

Shunt

Non-improve after Drainage

Follow

Protocol for NPH Patient

Clinical Evaluation (Outpatient)
(1) Symptom; Gait Disturbance, Memory Loss, Urinary Incontinence
(2) Radiological Finding; Head CT, MRI, Spinal MRI, Spectroscopy

Clinical Diagnosis of NPH

Three Days Drainage (Admission)

Improve after Drainage

Shunt

Non-improve after Drainage

**Two of Three Symptoms
with Ventriculomegaly**

Yes

Shunt

No

Follow

Responders to CSF drainage

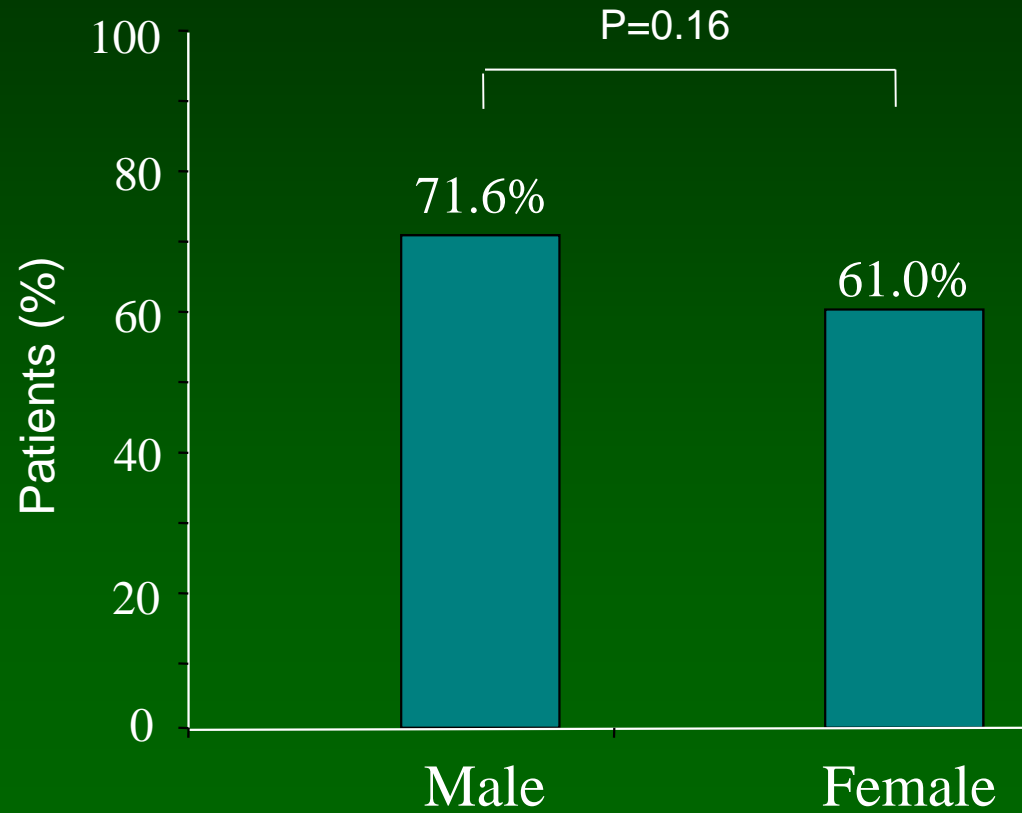
Only 2/3 of patients clinically diagnosed with idiopathic NPH will respond to CSF drainage

The remaining 1/3 will be exposed to risks of treatment with little or no benefit

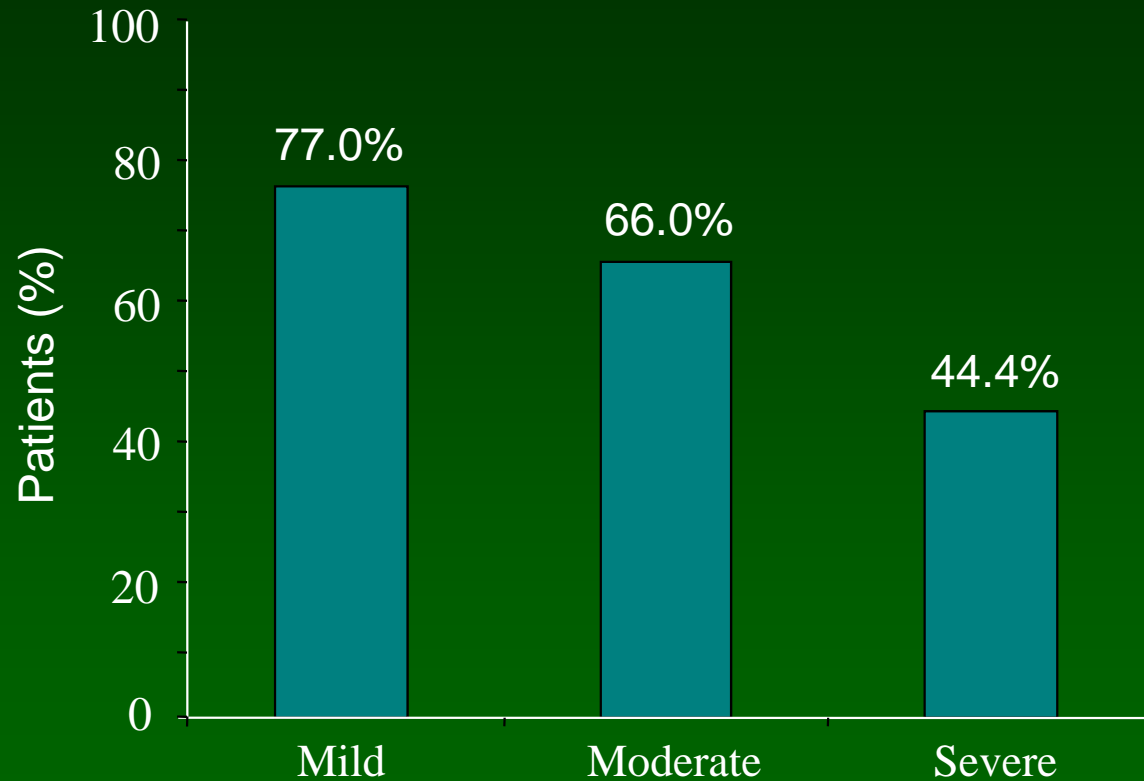
Age and Outcome in patients that improved following CSF drainage



Gender and Outcome that Improved following CSF Drainage

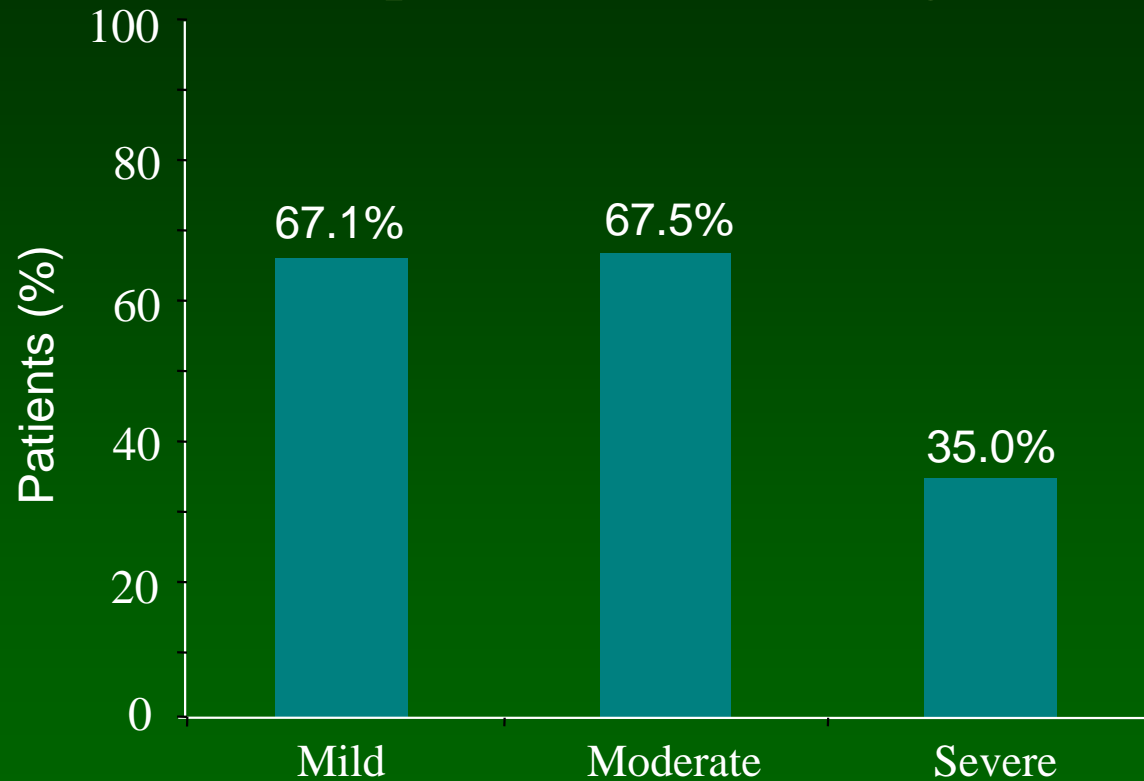


Pre-Drainage Symptom: Gait Improved with Drainage



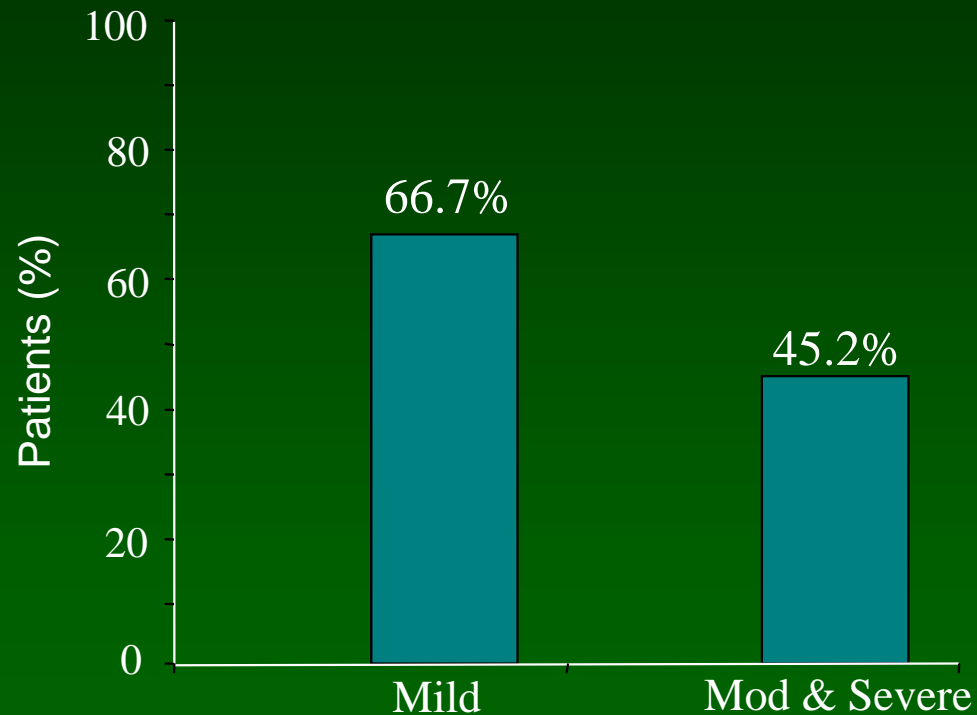
→ Patients who had mild and moderate gait disturbance significantly improved after drainage compared to severe ($p = 0.016$, *rank-sum test*).

Pre-Drainage Symptom: Memory Loss Improved with Drainage



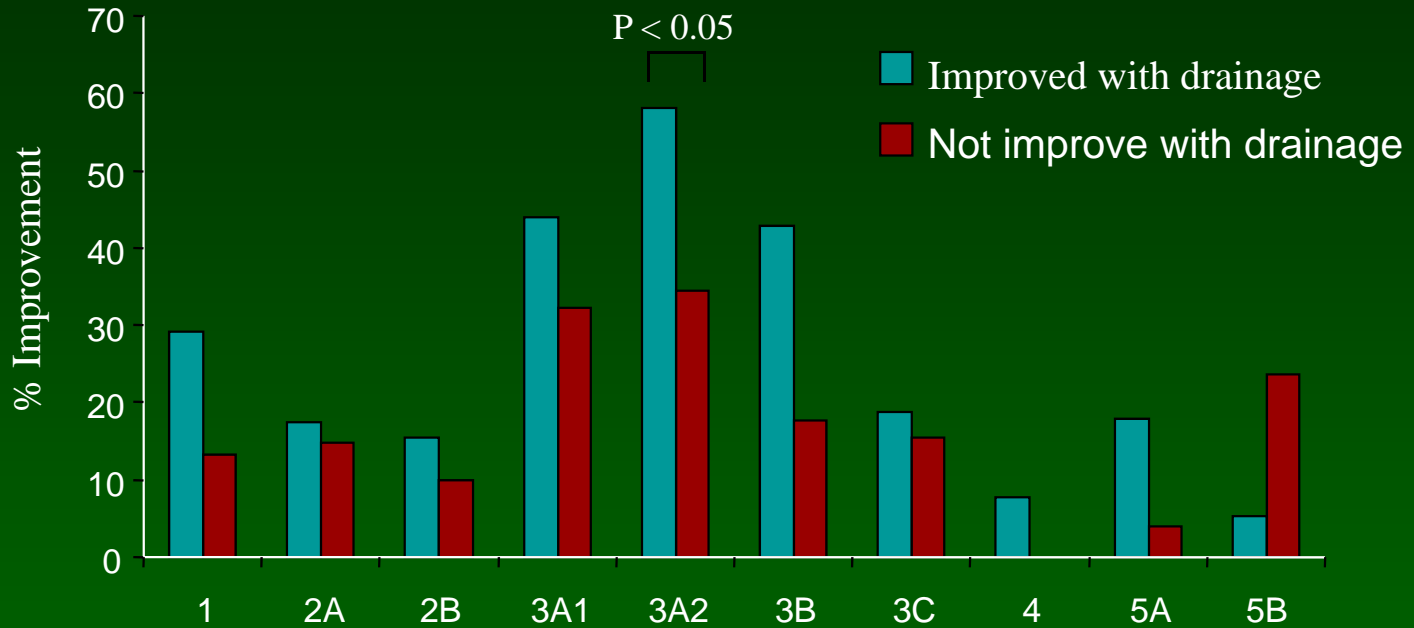
→ Patients who had mild and memory loss significantly improved after drainage compared to severe ($p = 0.0064$, *rank-sum test*).

Pre-Drainage Symptom: Incontinence Improved with Drainage



→ Patients who had mild urinary incontinence significantly improved after drainage compared to moderate and severe ($p = 0.034$, *rank-sum test*).

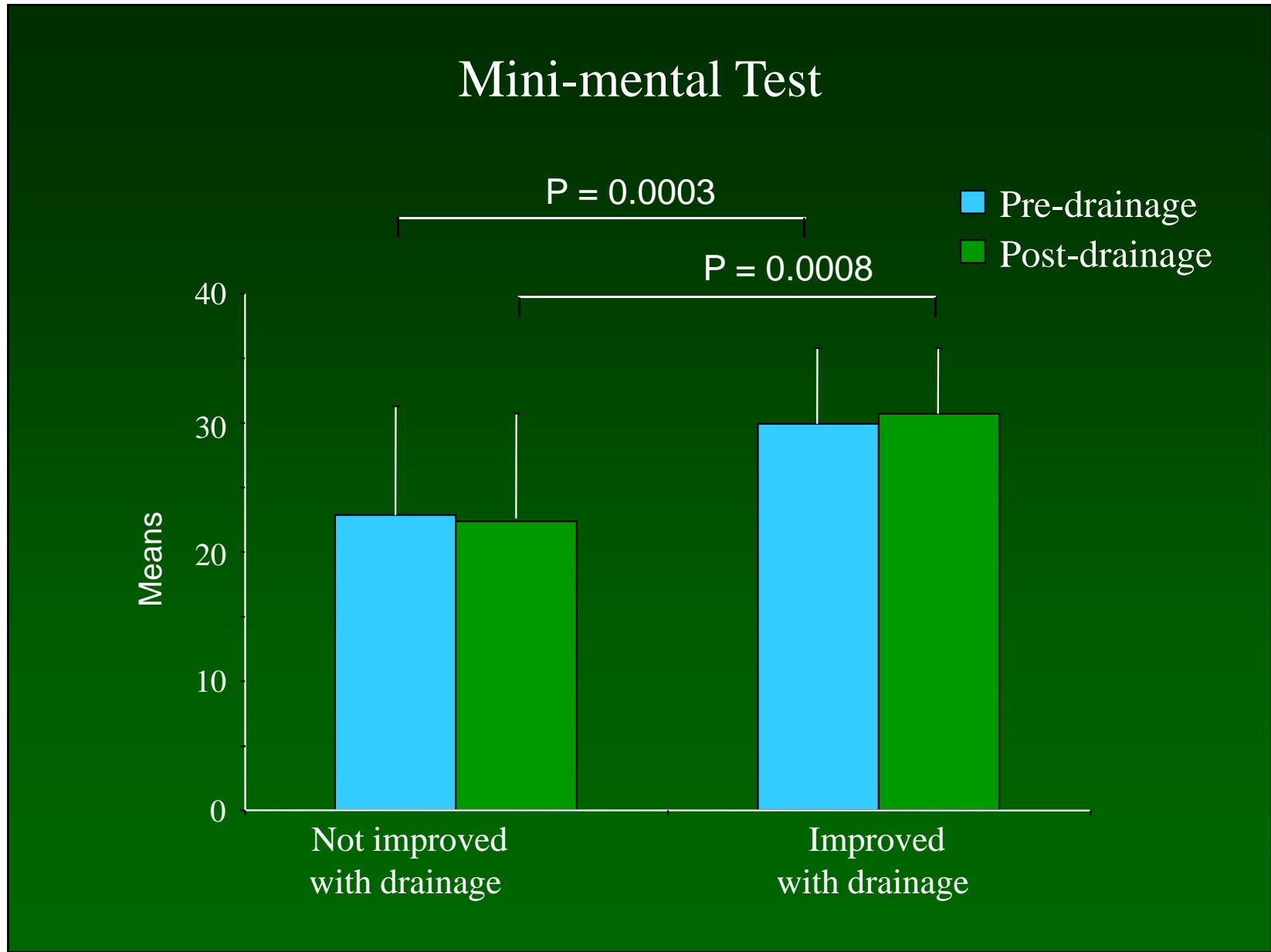
Neuropsychological Profile



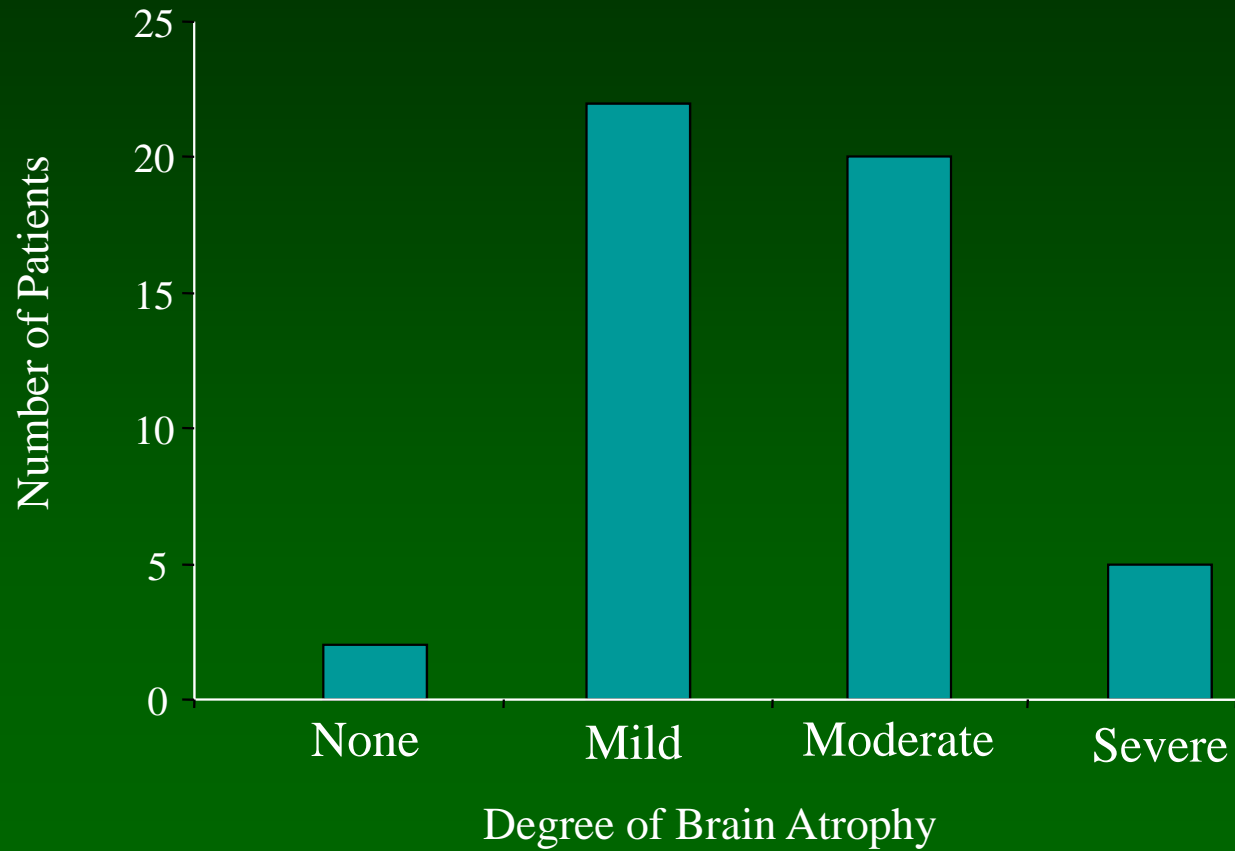
- 1. ATTENTION AND CONCENTRATION
- 2. LANGUAGE
 - A. WORD FINDING
 - B. AUDITORY COMPREHENSION

- 3. LEARNING AND MEMORY
 - A. AUDITORY MEMORY
 - 1. IMMEDIATE
 - 2. DELAYED
 - B. VISUAL MEMORY
 - C. LEARNING AUDITORY

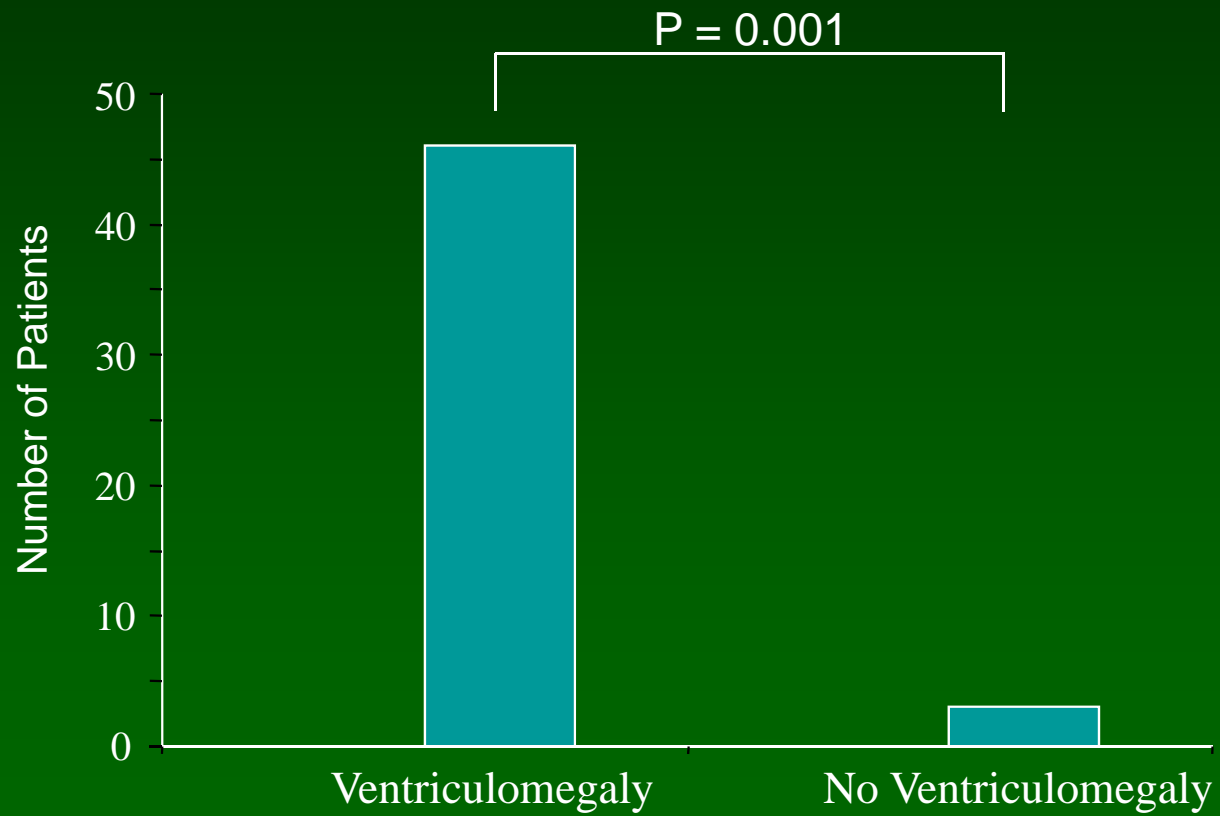
- 4. MOTOR
- 5. VISUAL SKILL
 - A. PERCEPTION
 - B. CONSTRUCTION



CT scan: Brain Atrophy in Patients Who Improved with Drainage



Ventriculomegaly and Outcome Improved with Drainage



Complications: ---

3 Day Drainage Protocol

2 Infections

Shunted Patients

3 SDH

3 Infections

3 double vision (transient)

4 Hearing loss (transient)

1 Hygroma

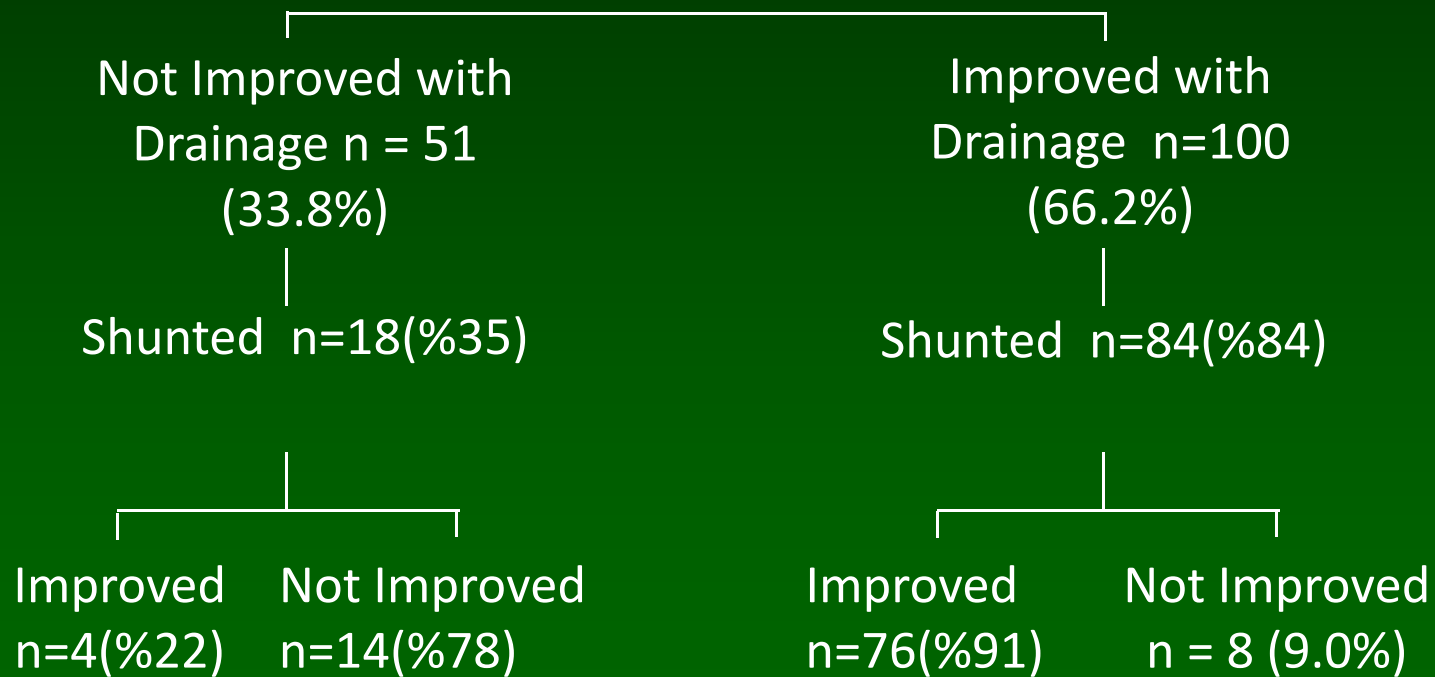
1 Death * (attempted biopsy)

Drainage Complication Rate 2/151 1.3%

Shunt Complication Rate 15/102 14.7%

Management of Normal Pressure Hydrocephalus

3 Day drainage (n = 151)



Conclusion

1. NPH patients should be diagnosed and treated by shunt in **early stage of NPH** (as early as possible), as NPH is a progressive disease.
2. For patients who improve with 3 day drainage, the probability of a positive shunt outcome is >90 %

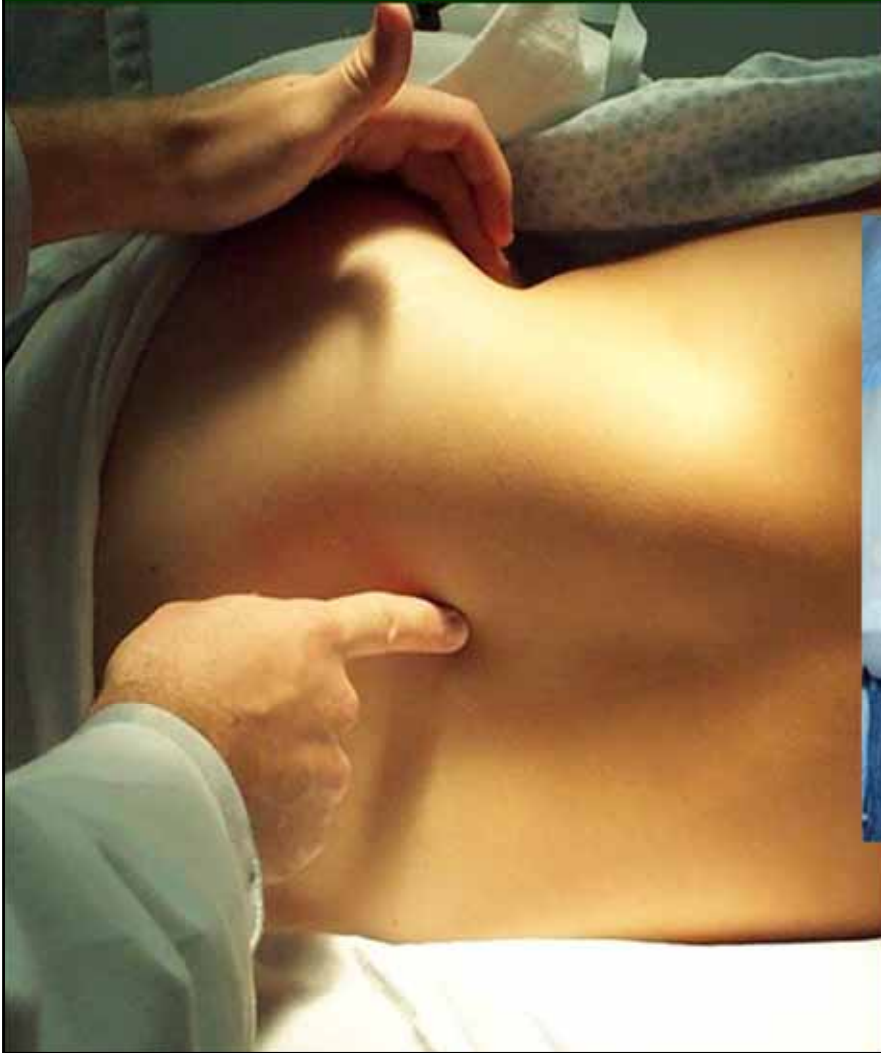
2. Definition of TAP 30/30

Observation of the Patient's gait at **30** minutes following a CSF removal of **30** cc

Hypotheses:

Patients improving at 30 minutes following drainage of 30 cc will identify the shunt responder

Lumbar Puncture



Protocol for NPH Patient

Clinical Evaluation (Outpatient)

- (1) Symptom; Gait Disturbance, Memory Loss, Urinary Incontinence
- (2) Radiological Finding; Head CT, MRI, Spinal MRI, Spectroscopy

Clinical Diagnosis of NPH

Tap 30 cc

Improve after Tap at 30 Min

Shunt
programmable
valve

Non-improve after Tap at 30 min

Follow

Protocol for NPH Patient

Clinical Evaluation (Outpatient)

- (1) Symptom; Gait Disturbance, Memory Loss, Urinary Incontinence
- (2) Radiological Finding; Head CT, MRI, Spinal MRI, Spectroscopy

Clinical Diagnosis of NPH

Tap 30 cc

Improve after Tap at 30 Min

Shunt with
programmable
valve

Non-improve after Tap at 30 min

Shunt with reduced expectation

Patients Profile

1) Total: n=107

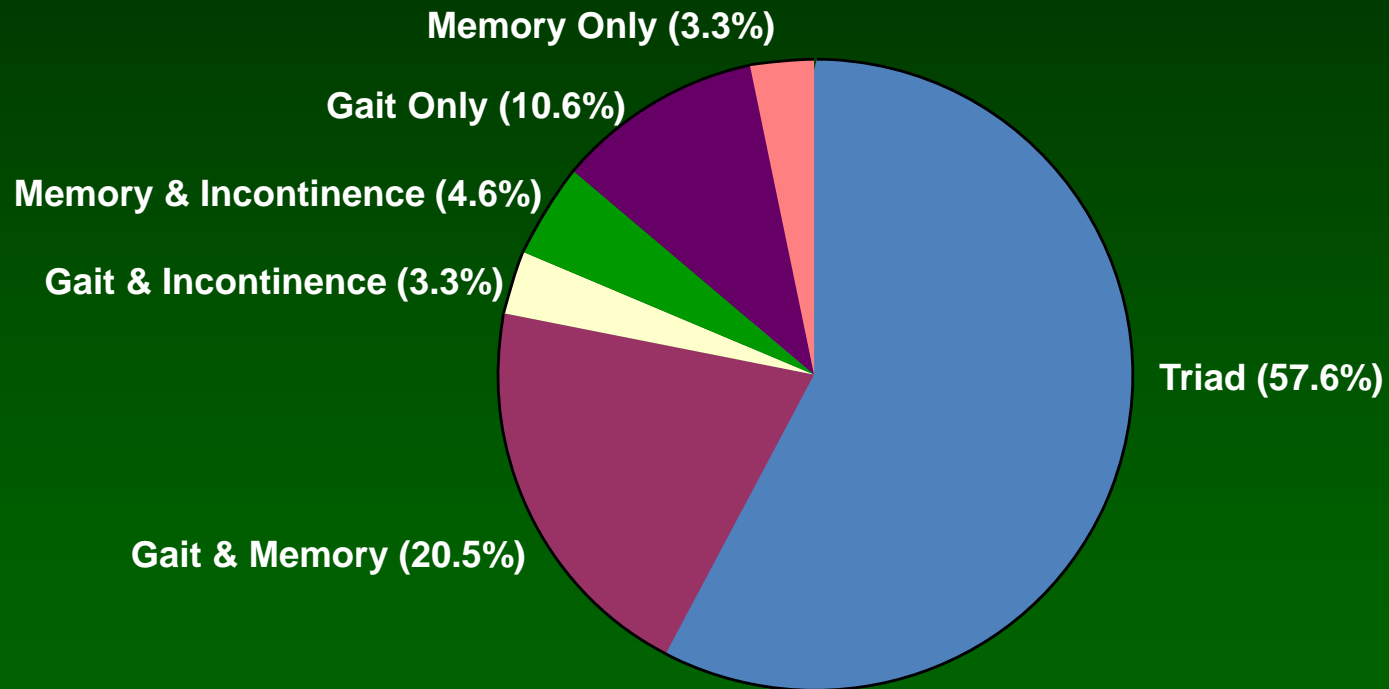
2) Age: 78.3 ± 7 yrs

3) Sex: Male n=66, Female n=41

Combination of Clinical Symptoms

- GAIT ONLY 6 %
- GAIT AND DEMENTIA 5 %
- GAIT AND INCONTINENCE 19 %
- GAIT, DEMENTIA, INCONTINENCE 70 %

Combination of Clinical Symptoms



Responders to TAP 30/30

- A total of 44 % of patients respond to TAP 30/30. The remainder (56 %) did not improve.
- Of those patients who improved with TAP 30/30, 100 % improved with Shunt.

Sensitivity of TAP 30/30

- Sensitivity 72 % 44/44 + 17
- Specificity 100 % 15/15 + 0
- Accuracy 78 %

Complications:

TAP 30/ 30

0 Infections

3 Severe Headaches

Shunted Patients (n=76)

3 SDH (acute)

3 Headache

1 double vision (transient)

2 Hearing loss (transient)

3 Hygroma

3 ICH

Tap 30/30 Complication Rate	3/107	1.3%	Recovery	100 %
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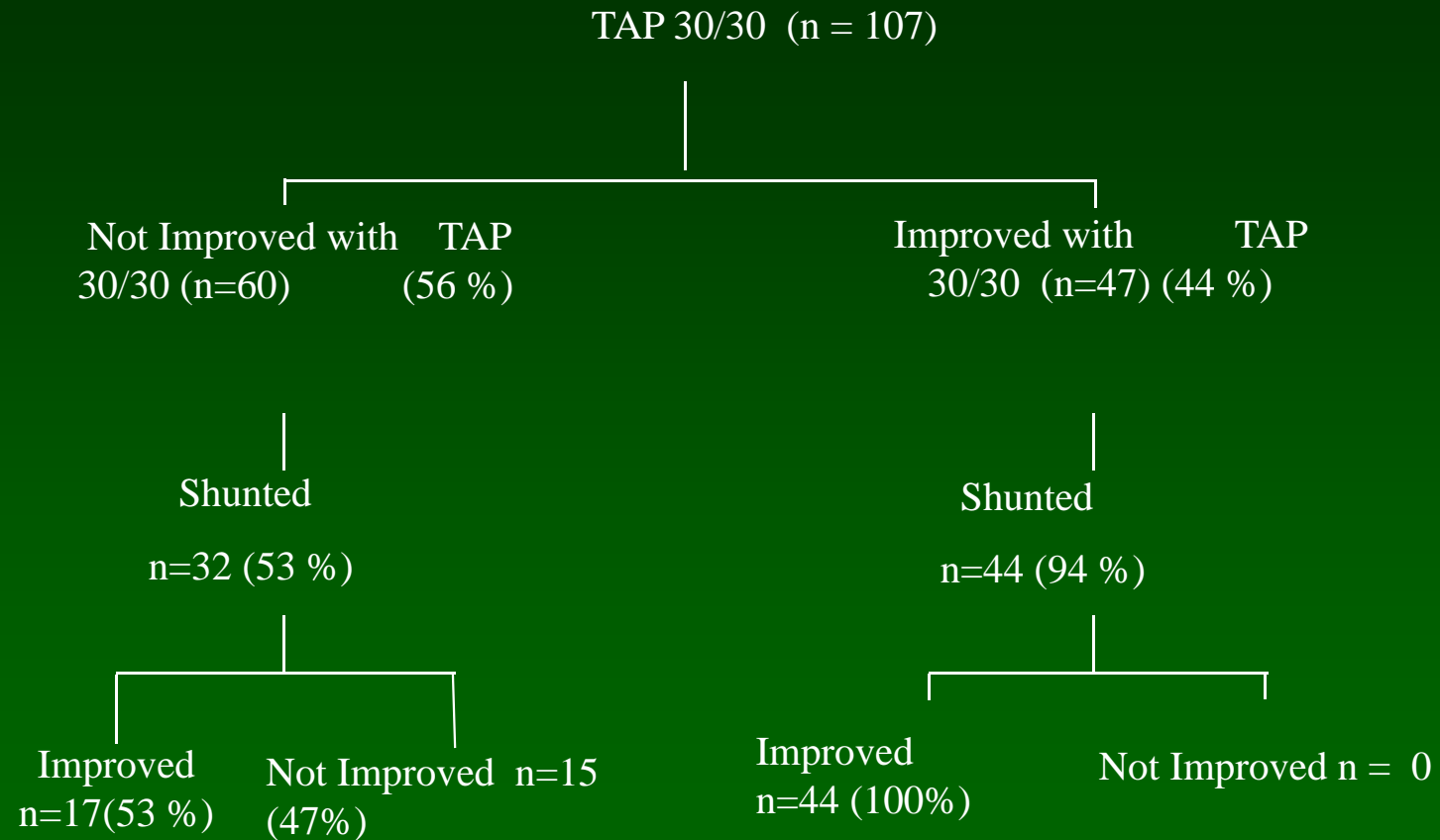
Shunt Complication Rate	15/76	19.7 %	Recovery	100 %
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Serious Shunt Complications	6/76	7.9 %	Recovery	100 %
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Conclusion:

1. NPH patients should be diagnosed and treated by shunt in early stage of NPH (as early as possible), as NPH is a progressive disease.
2. For patients who improve with TAP 30/30, the prediction of a positive shunt outcome is 100 %
3. The accuracy of TAP 30/30 to predict responders and non responders is 78 %

Management of Normal Pressure Hydrocephalus



Recommendation:

- If the patient responds to TAP 30/30 , proceed directly to shunt.
- If the patient is clinically diagnosed as INPH and does not respond to TAP 30/30, admit the patient for ELD.

Is there an upper age limit beyond which we should not recommend a shunt in this elderly population ?

See the Editorial and the Response in this issue, pp 971–973.

J Neurosurg 102:987–997, 2005

Diagnosis and management of idiopathic normal-pressure hydrocephalus: a prospective study in 151 patients

ANTHONY MARMAROU, PH.D., HAROLD F. YOUNG, M.D., GUNES A. AYGOK, M.D.,
SATOSHI SAWAUCHI, M.D., OSAMU TSUJI, M.D., TAKUJI YAMAMOTO, M.D.,
AND JANA DUNBAR, PH.D.

Department of Neurosurgery, Virginia Commonwealth University Medical Center, Richmond, Virginia

Object. The diagnosis and management of idiopathic normal-pressure hydrocephalus (NPH) remains controversial, particularly in selecting patients for shunt insertion. The use of clinical criteria coupled with imaging studies has limited effectiveness in predicting shunt success. The goal of this prospective study was to assess the usefulness of clinical criteria together with brain imaging studies, resistance testing, and external lumbar drainage (ELD) of cerebrospinal fluid (CSF) in determining which patients would most likely benefit from shunt surgery.

Methods. One hundred fifty-one patients considered at risk for idiopathic NPH were prospectively studied according to a fixed management protocol. The clinical criterion for idiopathic NPH included ventriculomegaly demonstrated on computerized tomography or magnetic resonance imaging studies combined with gait disturbance, incontinence, and dementia. Subsequently, all patients with a clinical diagnosis of idiopathic NPH underwent a lumbar tap for the measurement of CSF resistance. Following this procedure, patients were admitted to the hospital neurosurgical service for a 3-day ELD of CSF. Video assessment of gait and neuropsychological testing was conducted before and after drainage. A shunt procedure was then offered to patients who had experienced clinical improvement from ELD. Shunt outcome was assessed at 1 year postsurgery.

Conclusions. Data in this report affirm that gait improvement immediately following ELD is the best prognostic indicator of a positive shunt outcome, with an accuracy of prediction greater than 90%. Furthermore, bolus resistance testing is useful as a prognostic tool, does not require hospitalization, can be performed in an outpatient setting, and has an overall accuracy of 72% in predicting successful ELD outcome. Equally important is the finding that improvement with shunt surgery is independent of age up to the ninth decade of life in patients who improved on ELD.

KEY WORDS • idiopathic normal-pressure hydrocephalus •
normal-pressure hydrocephalus • hydrocephalus

STUDIES

Landmark study by Prof Anthony MARMAROU

151 patients prospectively studied according to a fixed management protocol.

- **Gait improvement** immediately followed ELD is the best prognostic indicator of a positive shunt outcome.
- Improvement with shunt surgery is independent of age up to the **ninth decade** of life in patient who improved on ELD.

Comparison of ELD and TAP 30/30 to Predict Shunt Outcome

Sensitivity: The percentage of patients with positive response to CSF drainage among total improved shunt outcomes

ELD = 0.95 TAP = 0.72

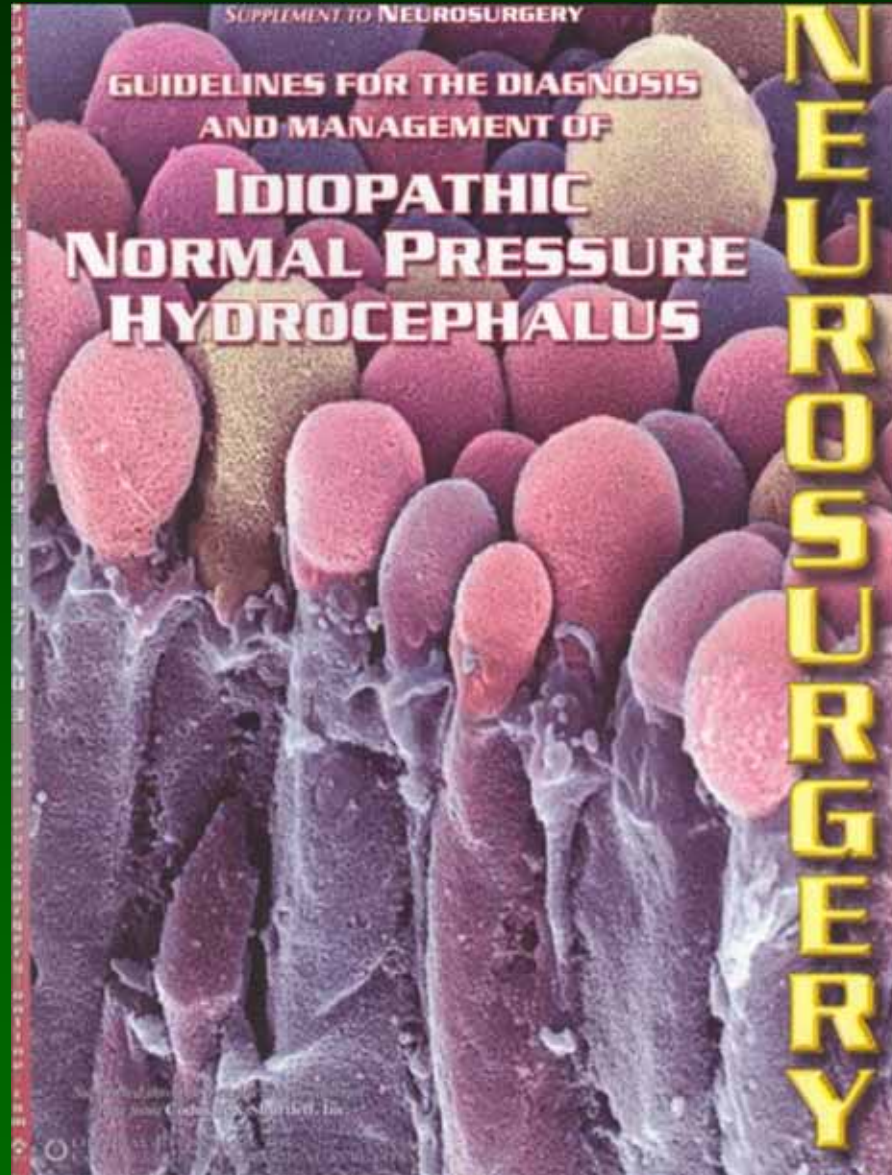
Specificity: The percentage of patients with poor response to CSF drainage outcomes among total poor shunt outcomes

ELD = 0.64 TAP = 100 % (no false positives)

Accuracy: % of Total Correct Predictions

ELD = 89.3 % TAP = 78 %

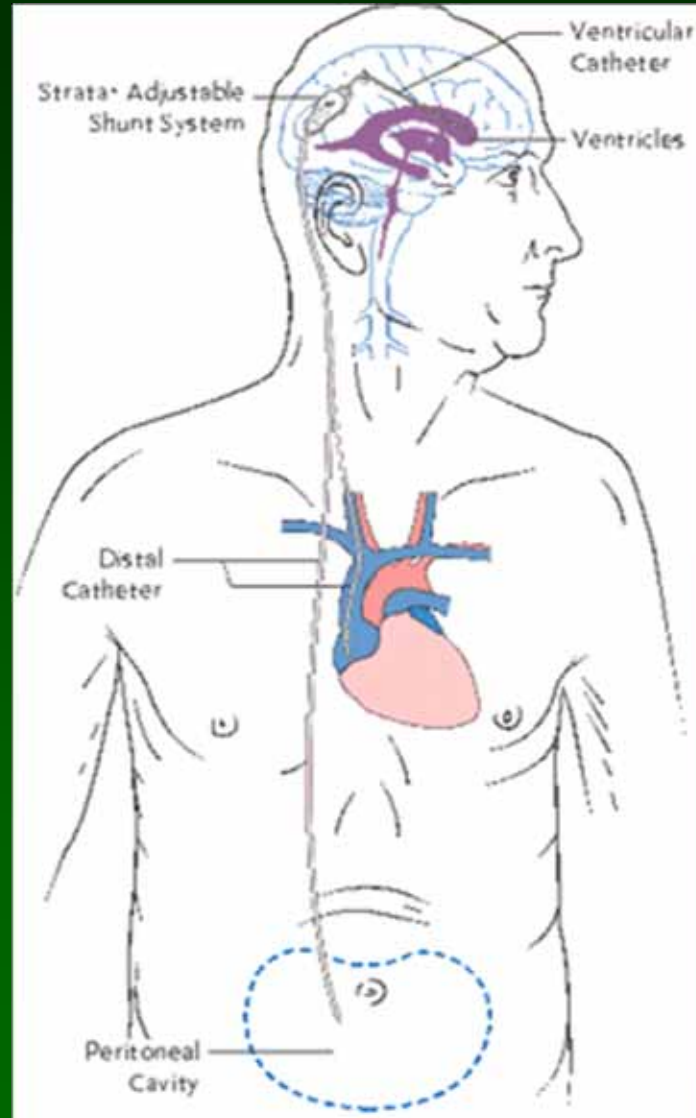
How do these results effect
guidelines for management ?

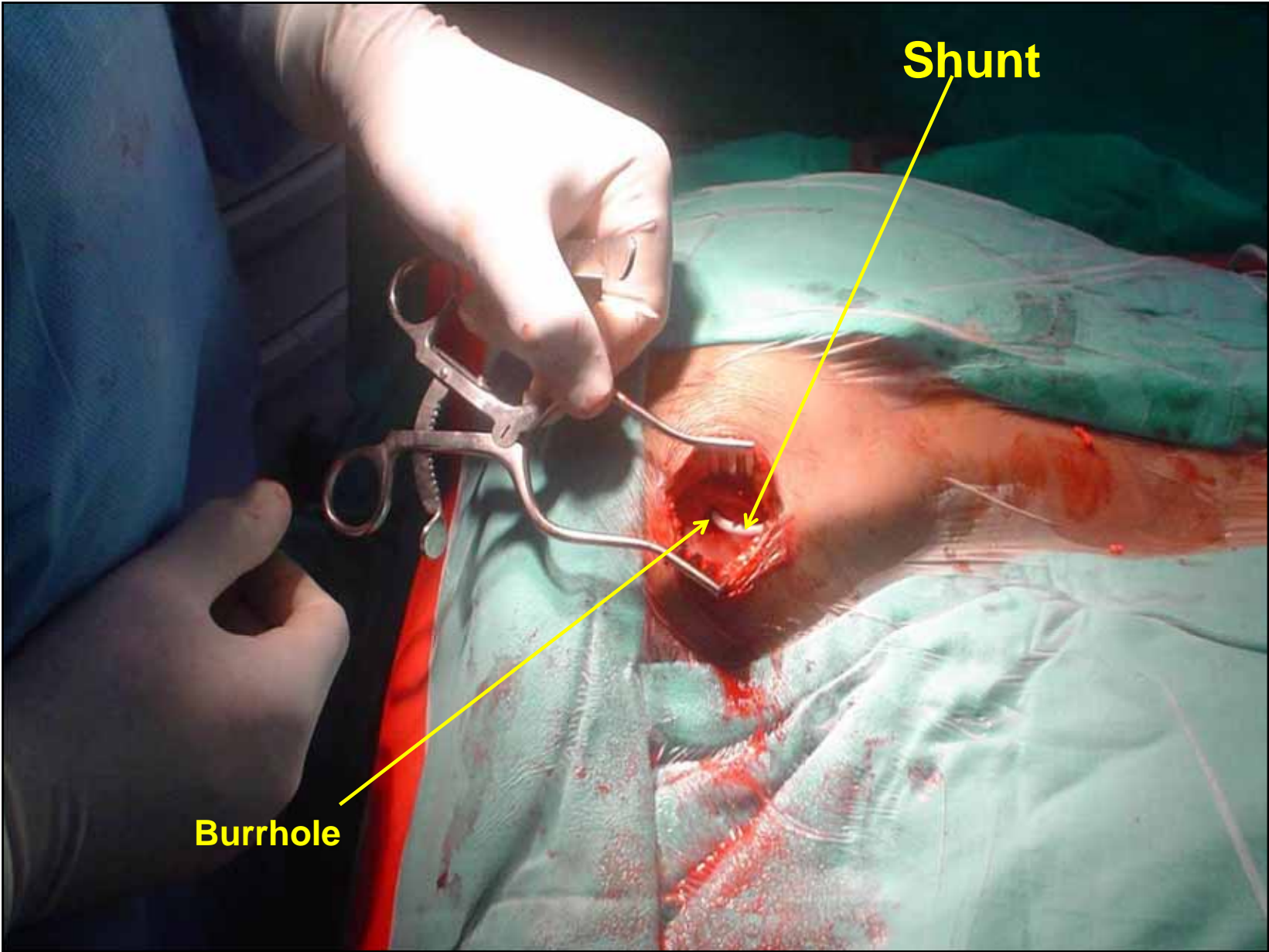


Sept 2005

Treatment Options

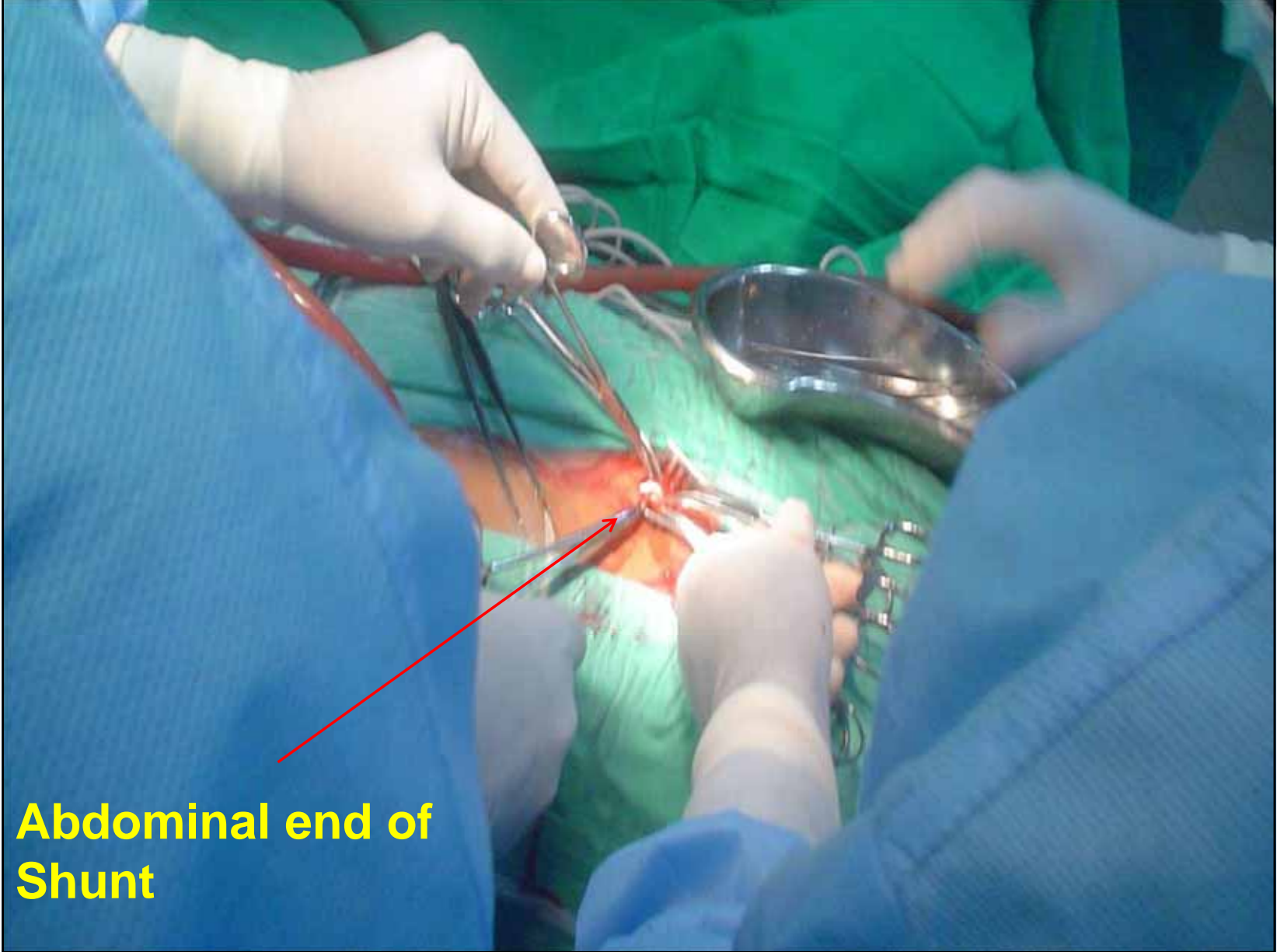
VPS (Ventriculo- peritoneal Shunt)





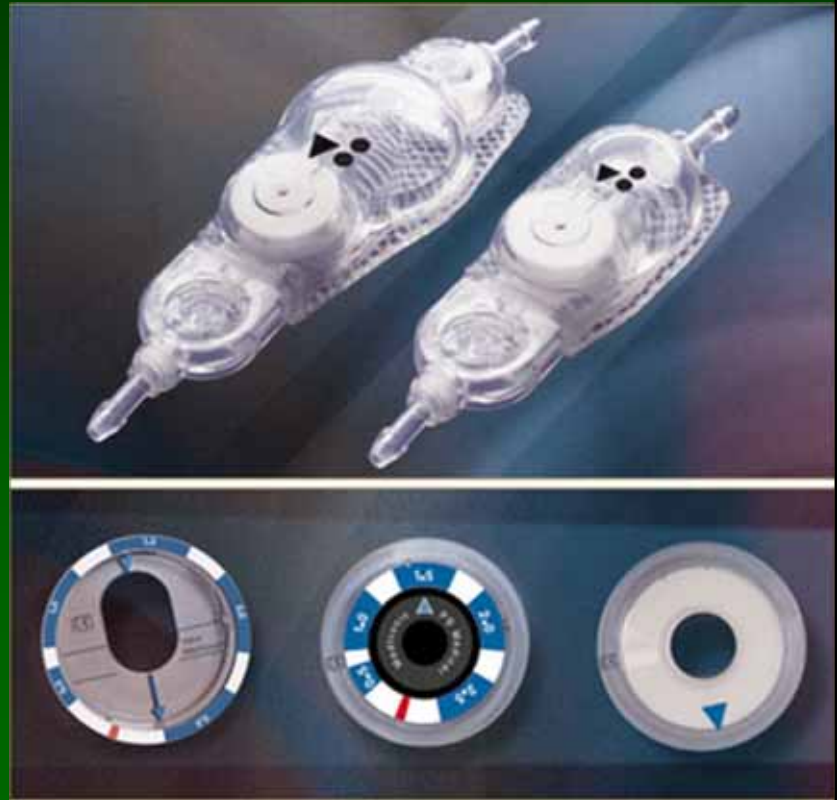


Tunneling of shunt



Abdominal end of Shunt

Programmable Shunts



Shunt

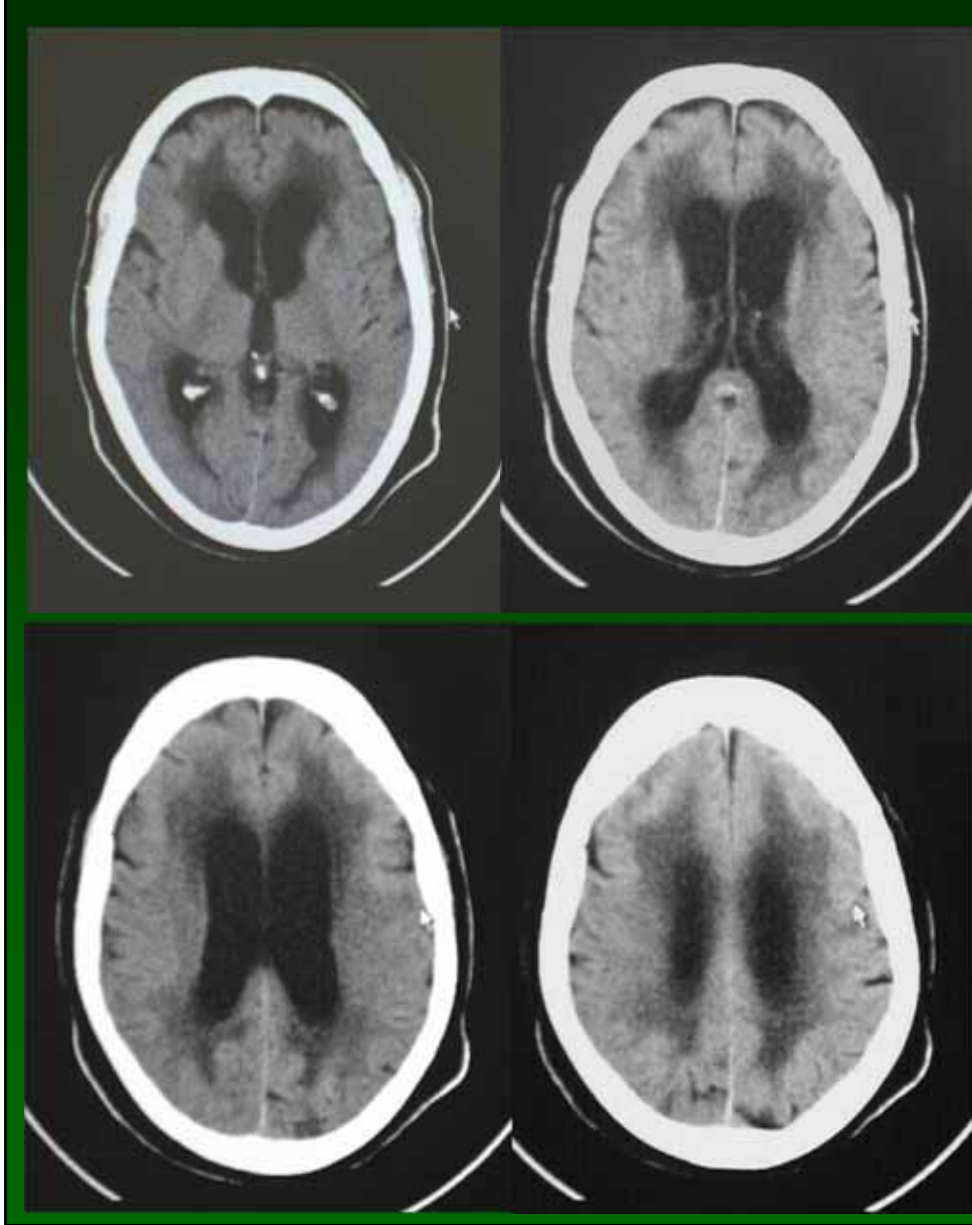


Programmable Shunts



Re-programming the shunt

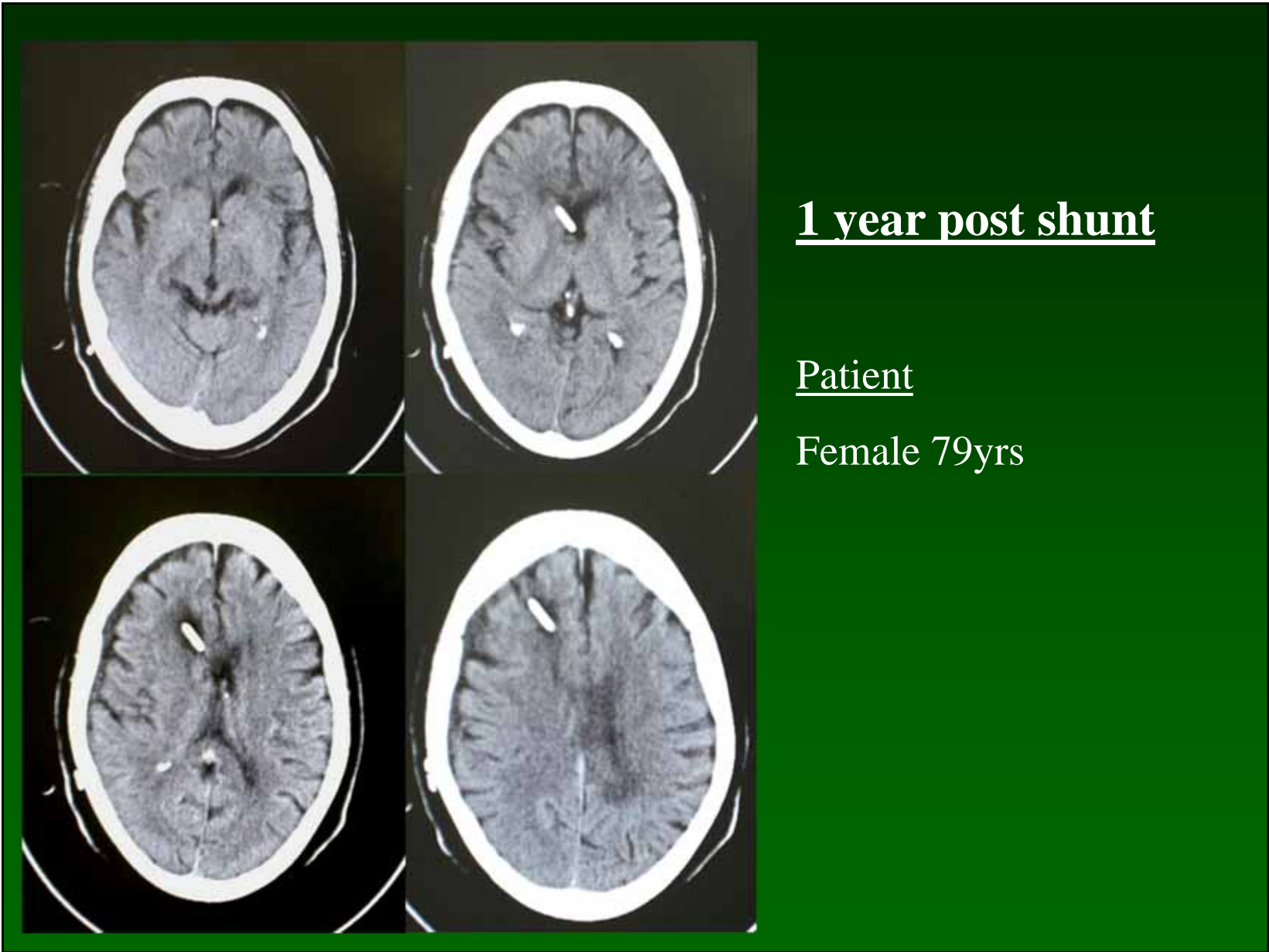




Before Shunt

Patient

Female/79yrs

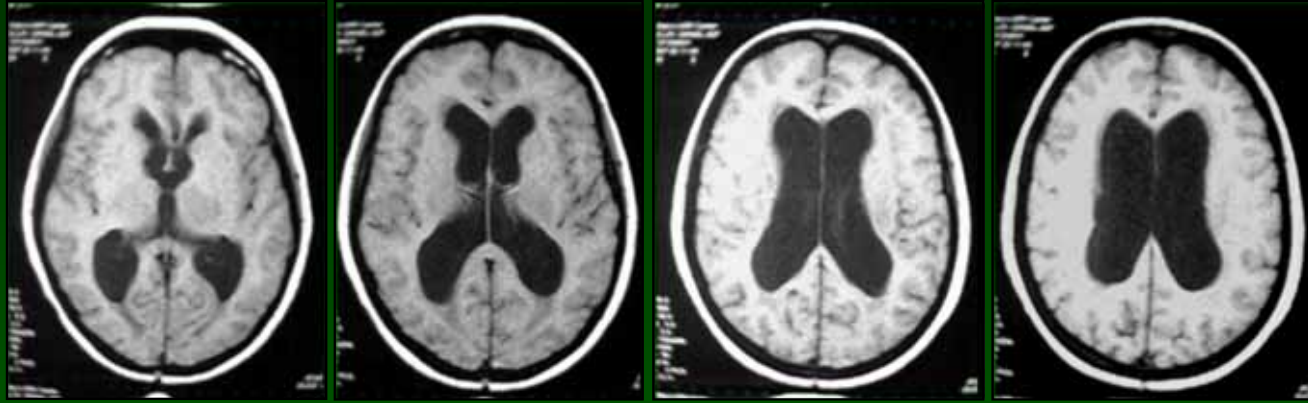


1 year post shunt

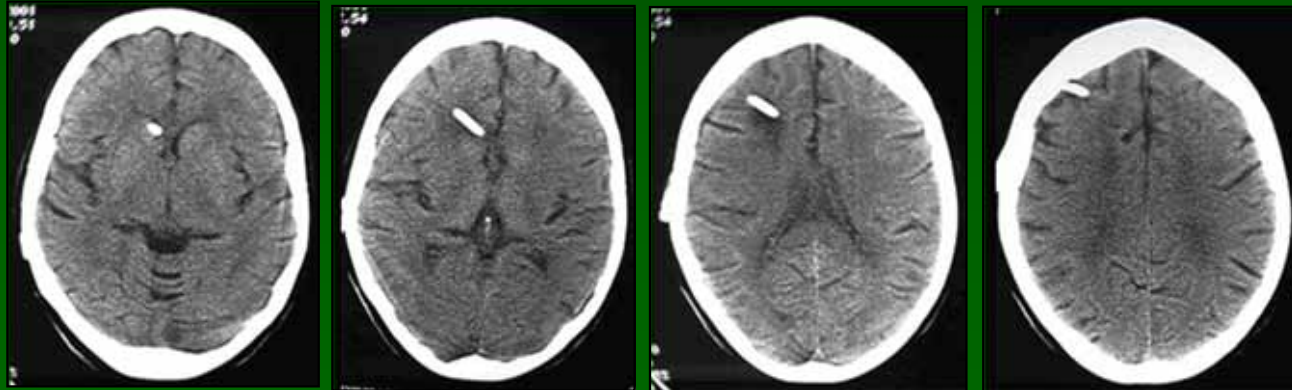
Patient

Female 79yrs

Pre Shunt



11 Months Post Shunt (V-P Programmable)



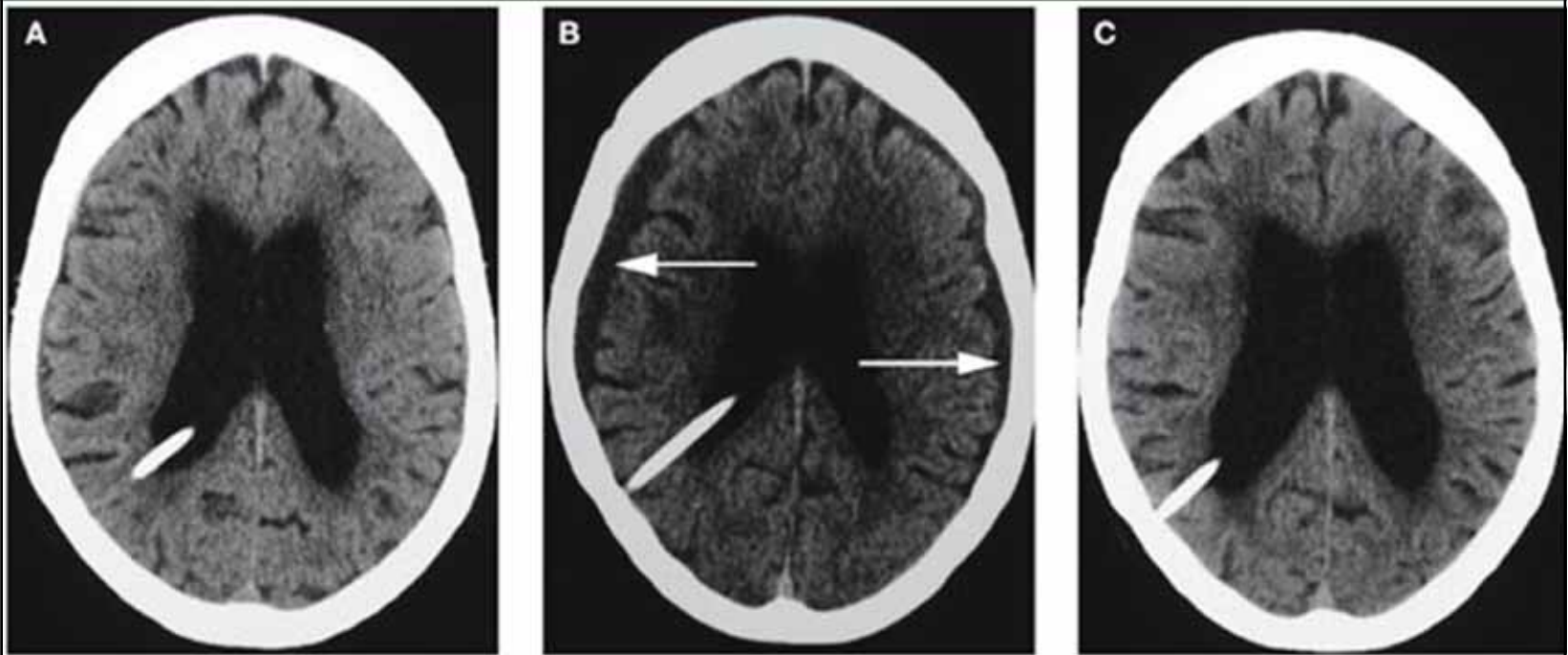
Severe Complications of Surgery Range from 5 to 35 %

- Infection
- Subdural Hematoma
- Hygroma
- Death

Percent Complications Related to the Shunt

Authors	%
Stein <i>et al</i> (1974)	15
Hughes <i>et al</i> (1978)	35
Black (1980)	38
Peterson <i>et al</i> (1985)	31
Spanu <i>et al</i> (1986)	13
Kosteljanetz <i>et al</i> (1990)	16
Larsson <i>et al</i> (1991)	50
Vannest <i>et al</i> (1992)	28
Marmarou <i>et al.</i> (2005)	13

Over Drainage



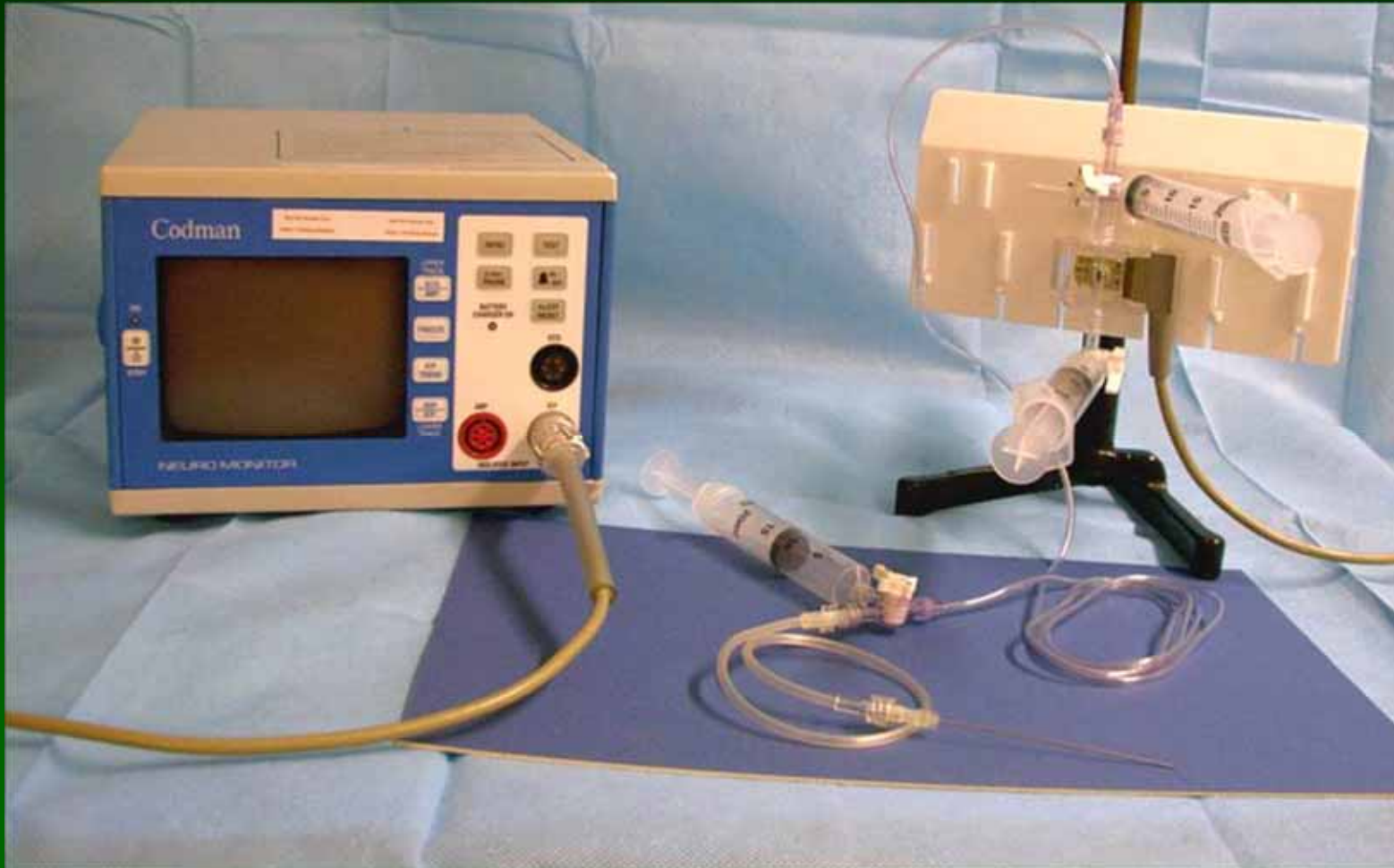
Before Shunting



After Shunting

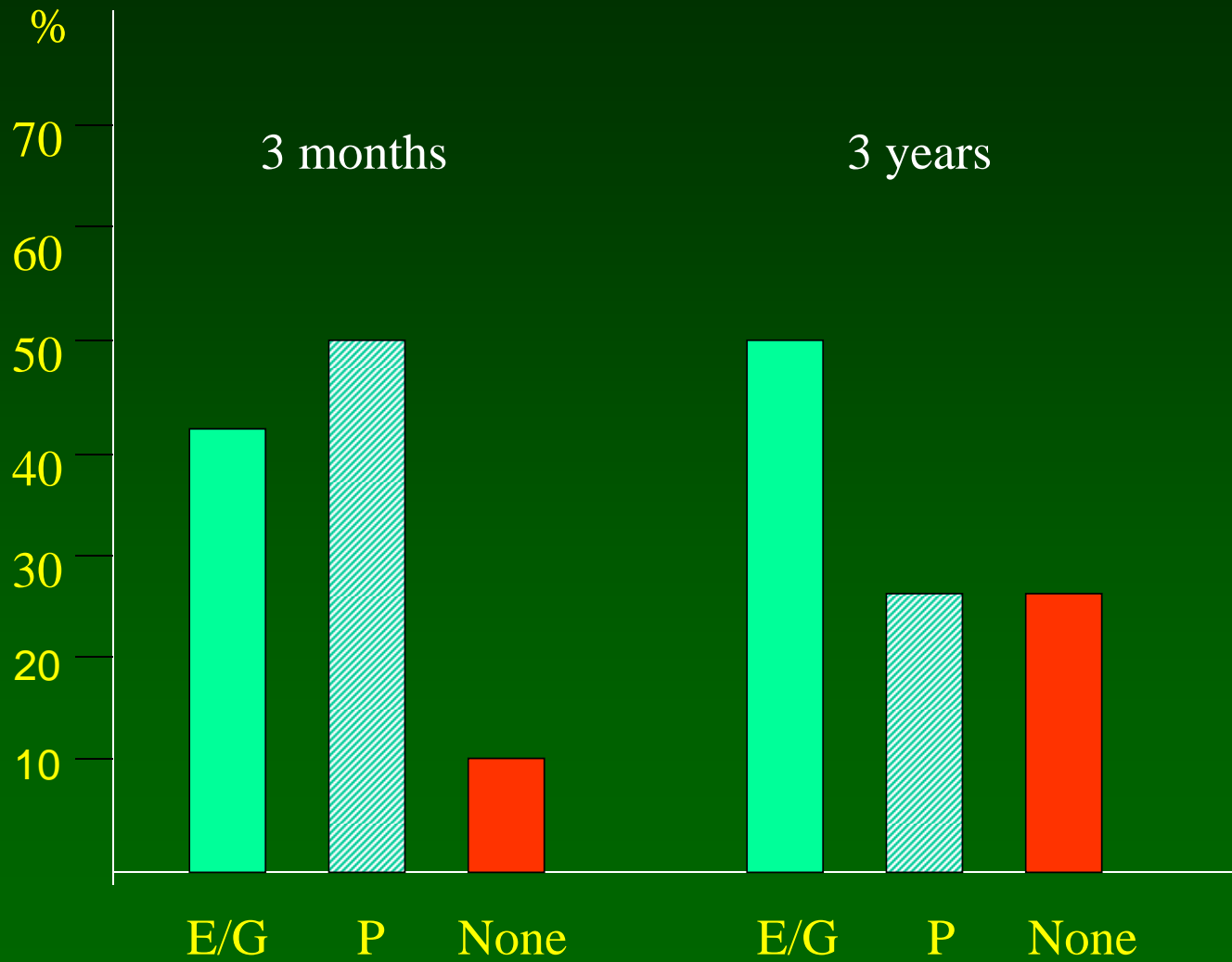


Lumbar Drainage Equipment

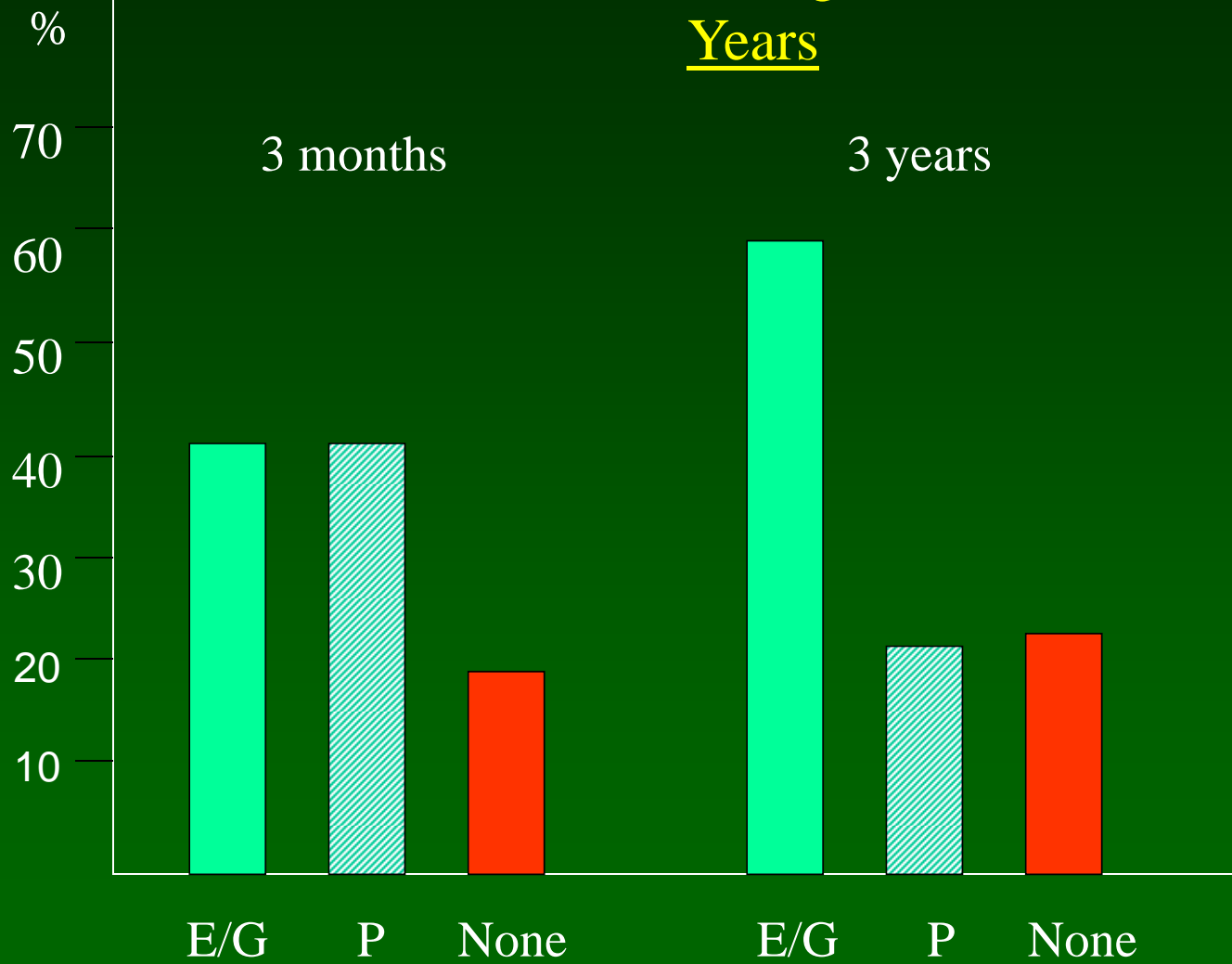


OUTCOME LONG TERM ?

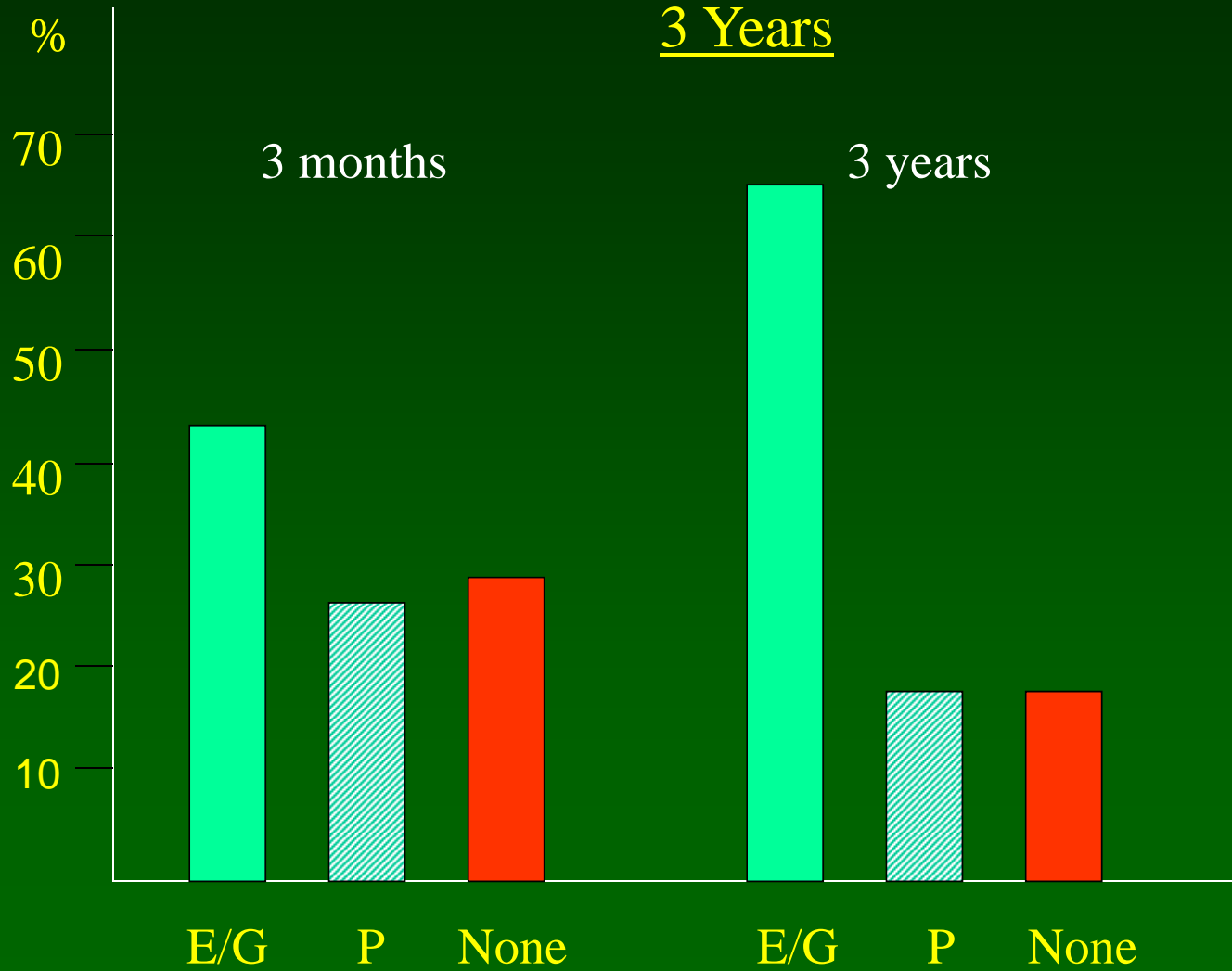
Gait Outcome Following Shunt : 3 Months vs 3 Years



Dementia Outcome Following Shunt : 3 Months vs 3 Years



Incontinence Outcome Following Shunt : 3 Months vs 3 Years



Because NPH is treatable, it is the one cause of “Dementia” that is considered reversible

NPH should be included in all diagnostic algorithm of Dementia

- ❖ NPH is a true diagnostic challenge but a rewarding one
- ❖ Keep a high index of suspicion
- ❖ Advent of programmable shunt for NPH is a major advance

*There are no bad patients
only
bad doctors!!*



Some other common causes of surgically reversible dementia

- Chronic Subdural Haematoma
- Brain Tumors
- Traumatic Brain Injury

CSDH

Reversible Dementia in patients with chronic subdural haematoma

Dementia is reversible in many patients with CSDH and surgery can improve not only independence in ADL (activities of daily living) but also neuropsychiatric functions.

HDS – R : Hasegawa Dementia Scale

MMSE : Revised Mini Mental State Examination

Chronic Subdural Haematoma

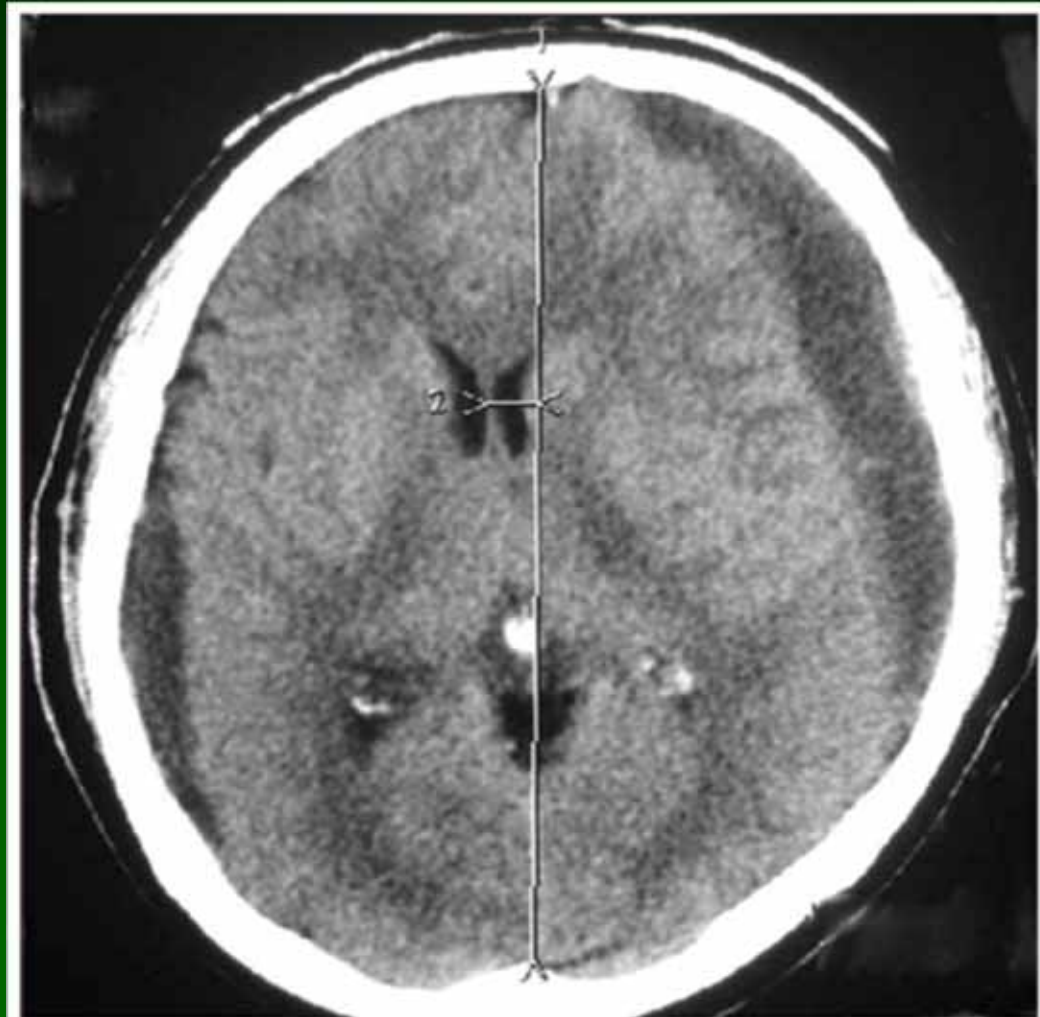
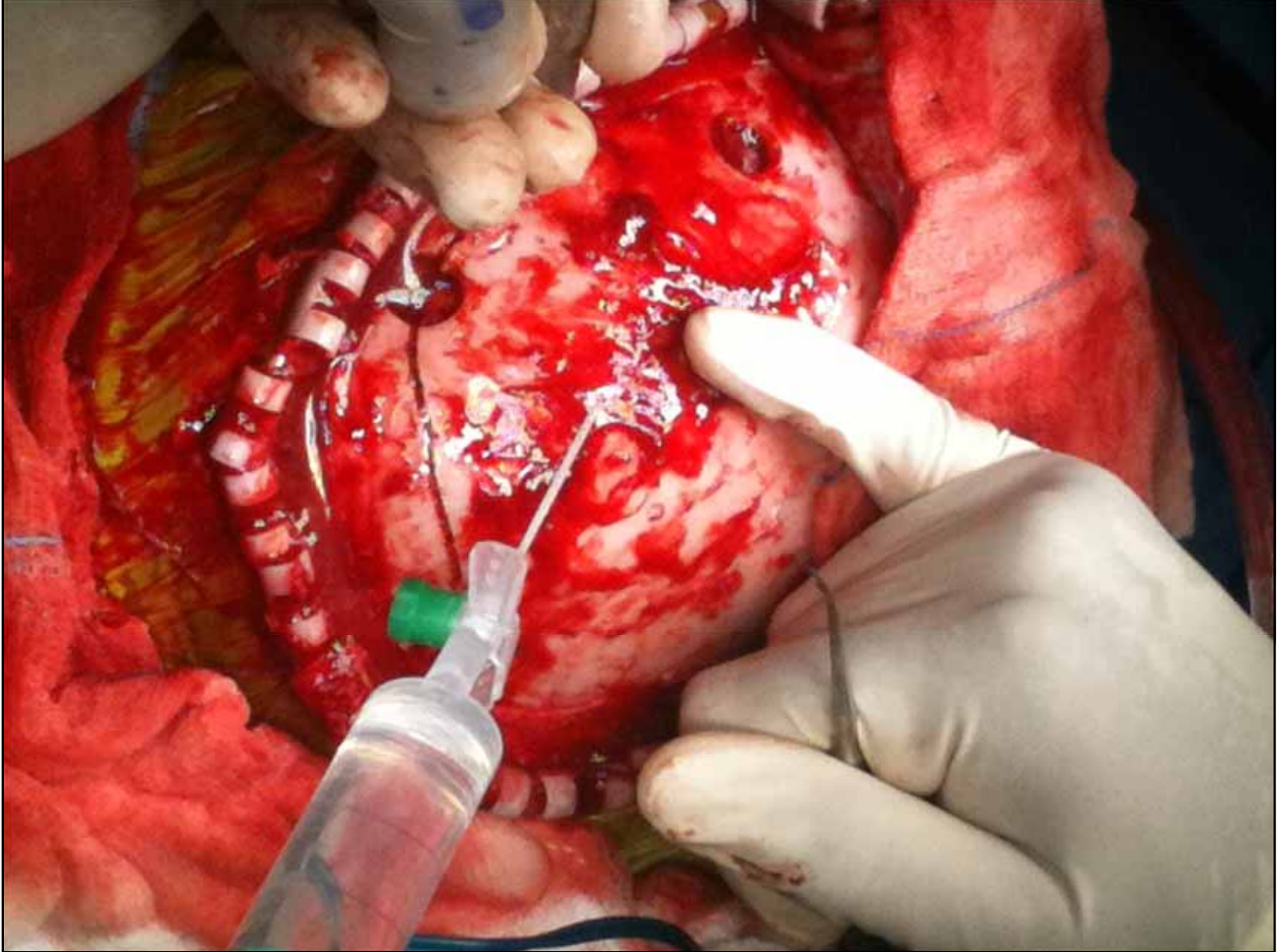
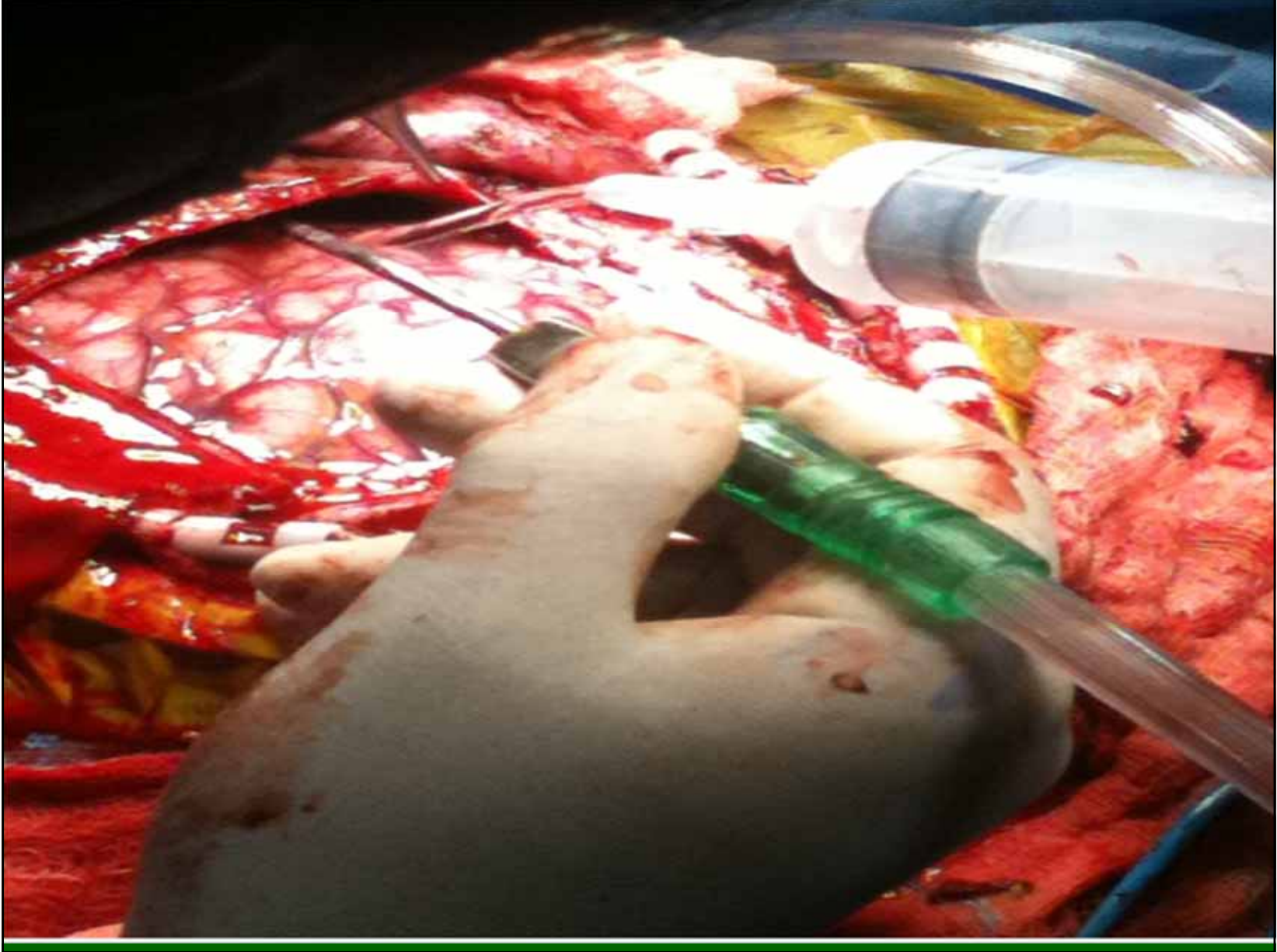
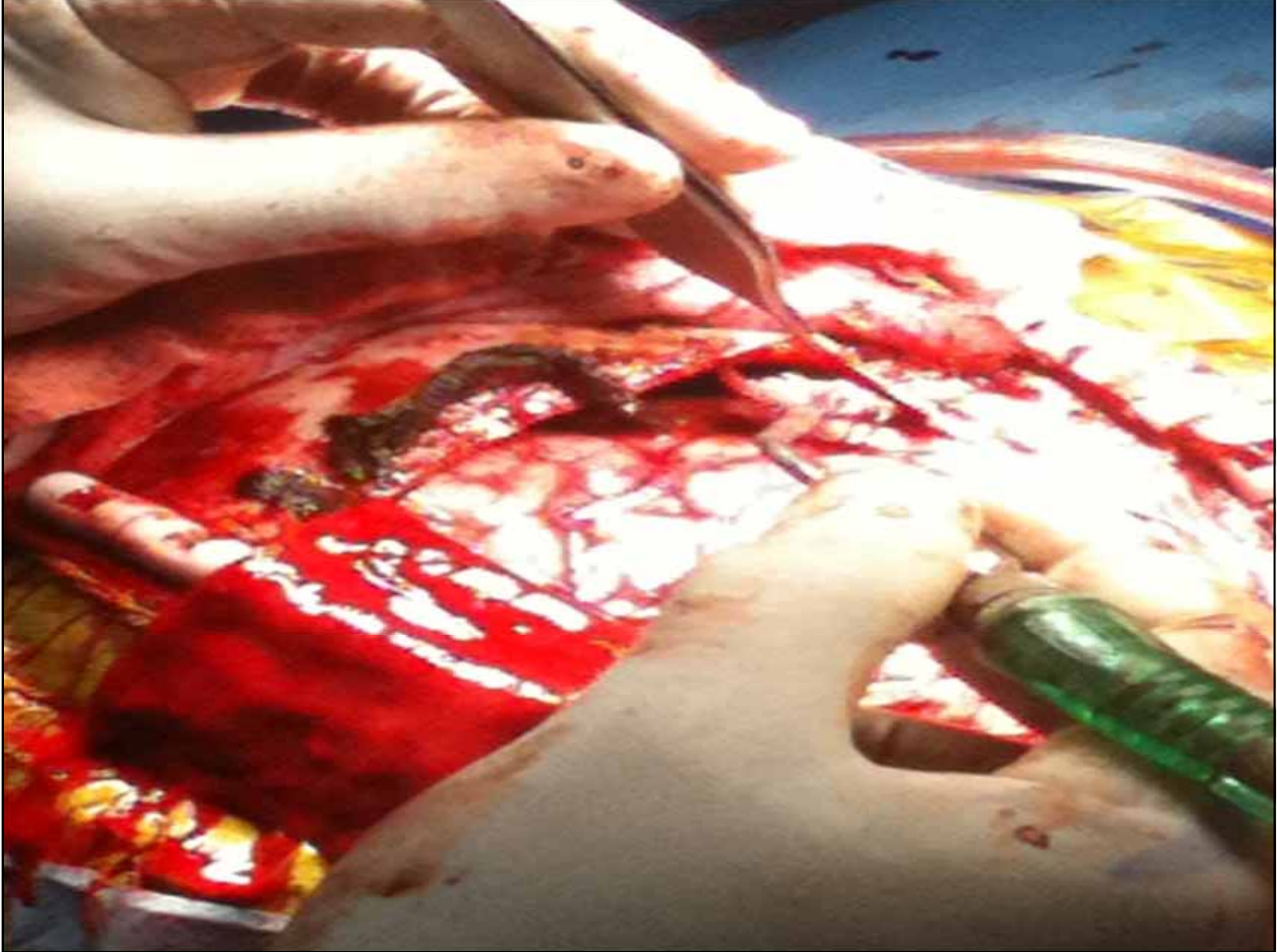


Fig 1: CT scan showing bilateral subdural collections, larger on the left.

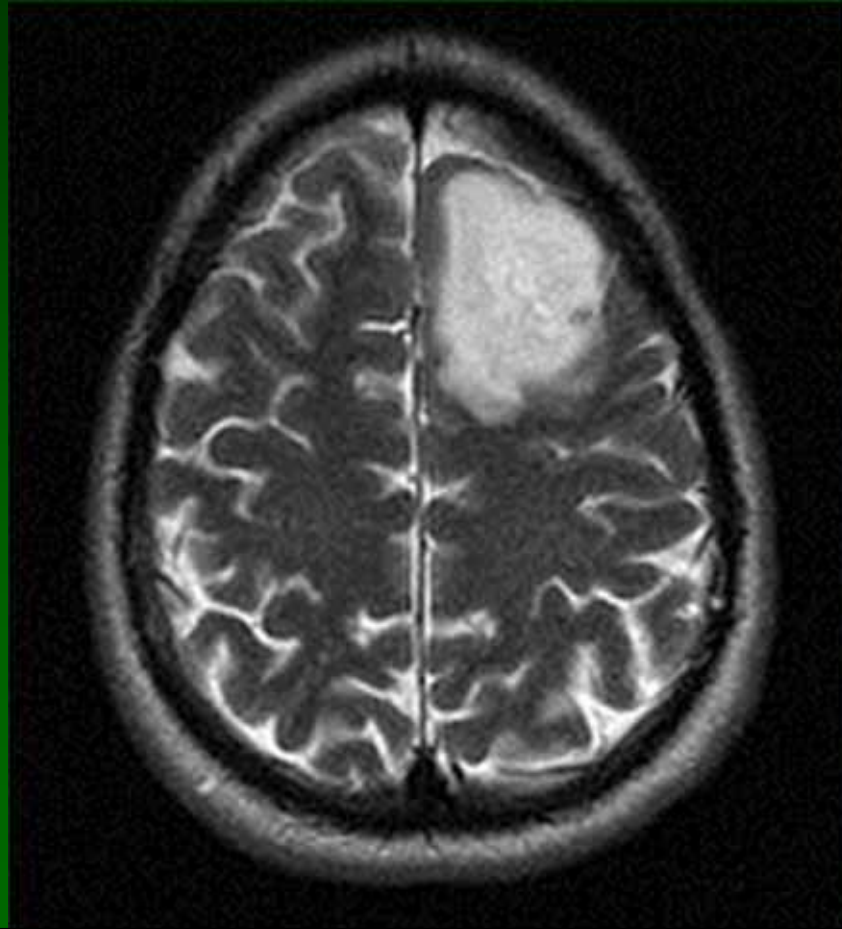




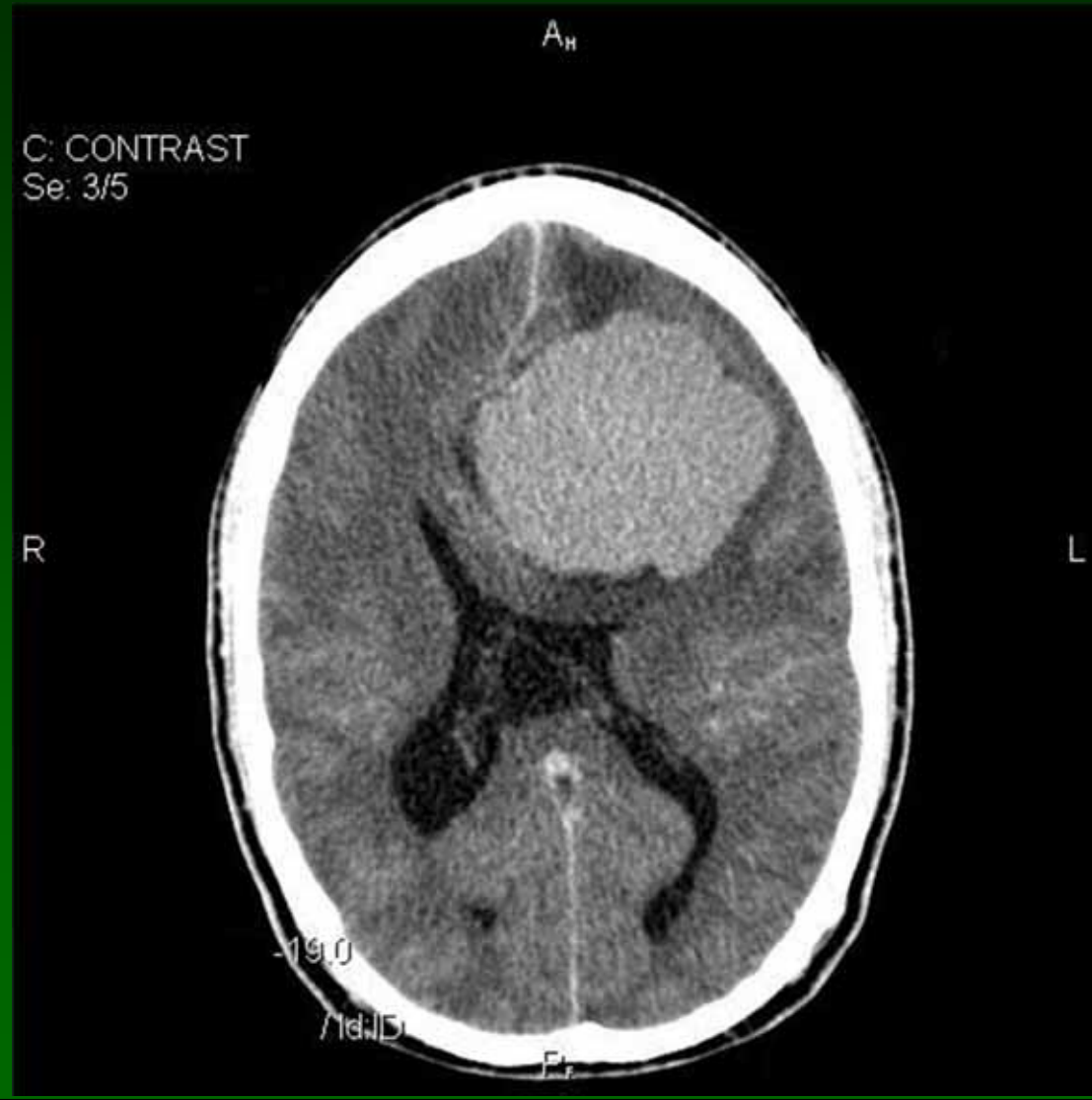




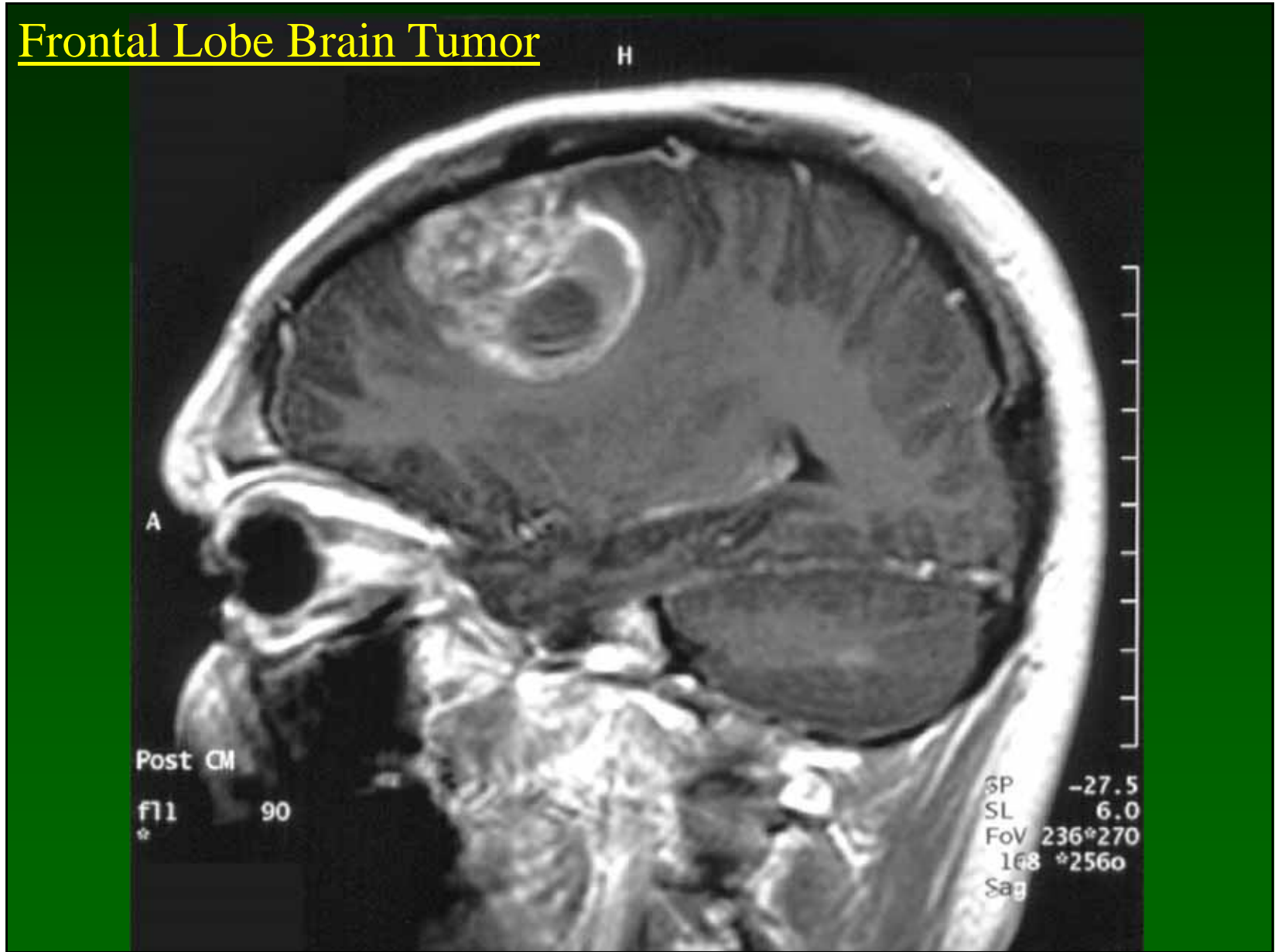
Left Frontal Lobe Brain Tumor



Left Frontal Lobe Brain Tumor



Frontal Lobe Brain Tumor



*Dementia should no longer
be an accepted side effect of
growing old.*



*We live our life with
passion...*

*We must practice our craft
with a passion greater than
life itself...*