NORMAL PRESSURE HYDROCEPHALUS(NPH)

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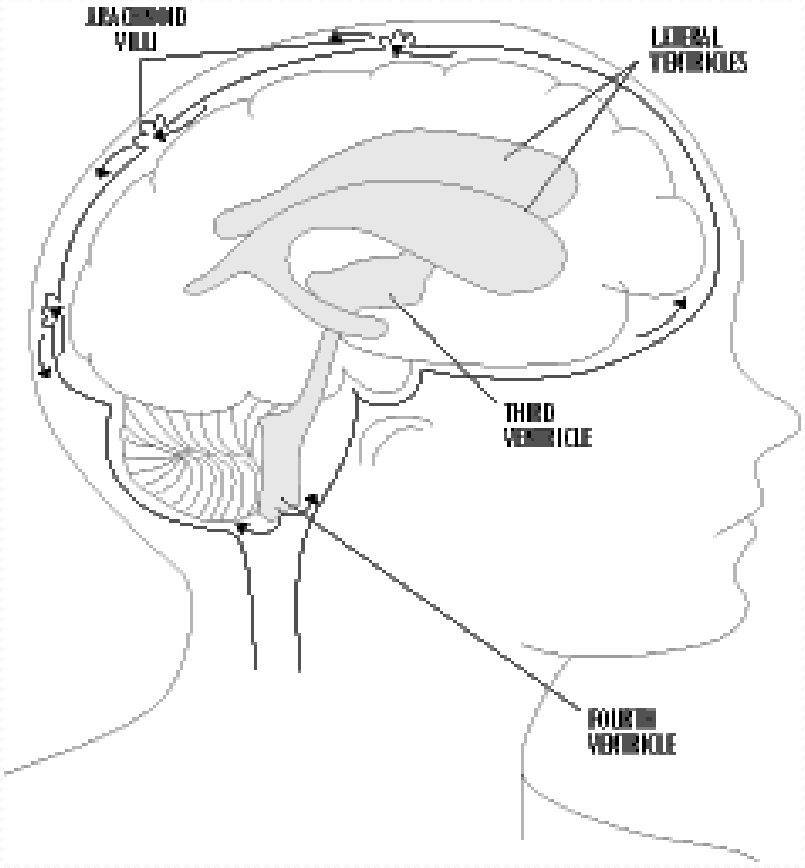
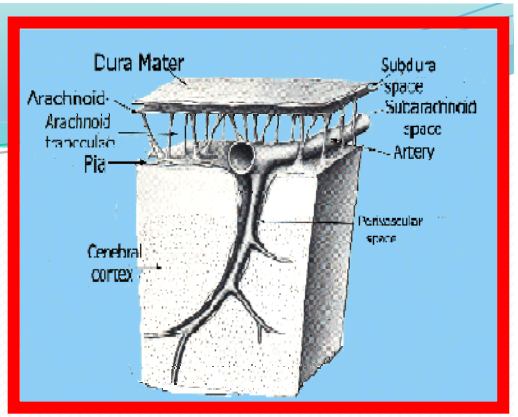
FIRST YEAR PG
DEPT OF GENERAL MEDICINE

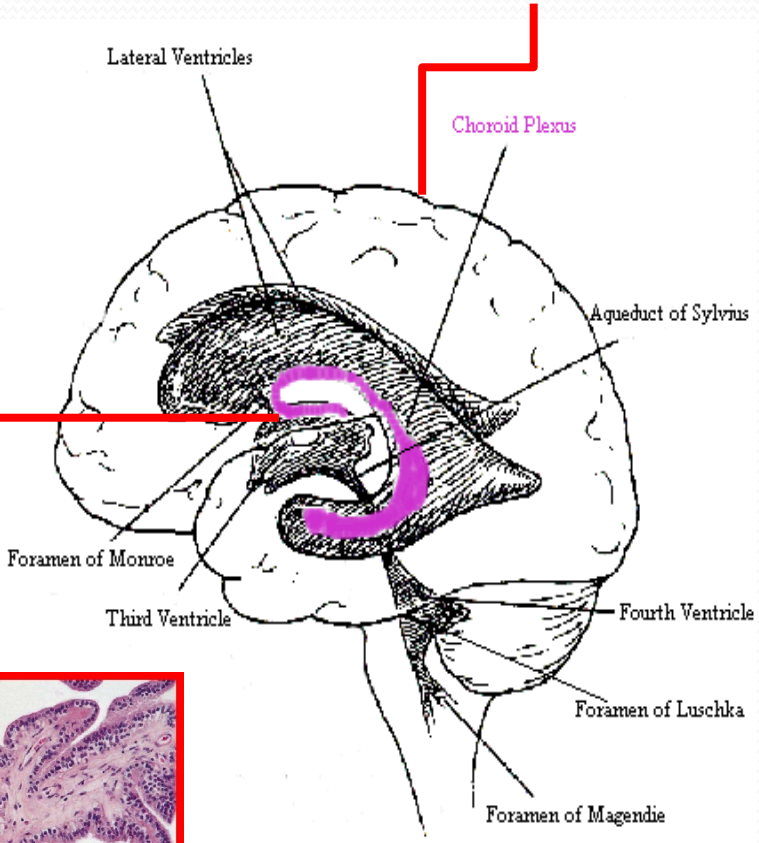
Overview of CSF production

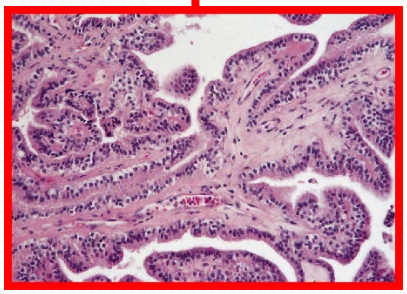
- The CSF volume of an average adult ranges from 80 to 160 ml
- The ventricular system holds approximately 20 to 50 ml of CSF
- CSF is produced in the choroid plexuses at a daily rate of 14-36 ml/hr



- The choroid plexuses are the source of approximately 80% of the CSF
- The blood vessels in the subependymal regions, and pia also contribute to the formation of CSF

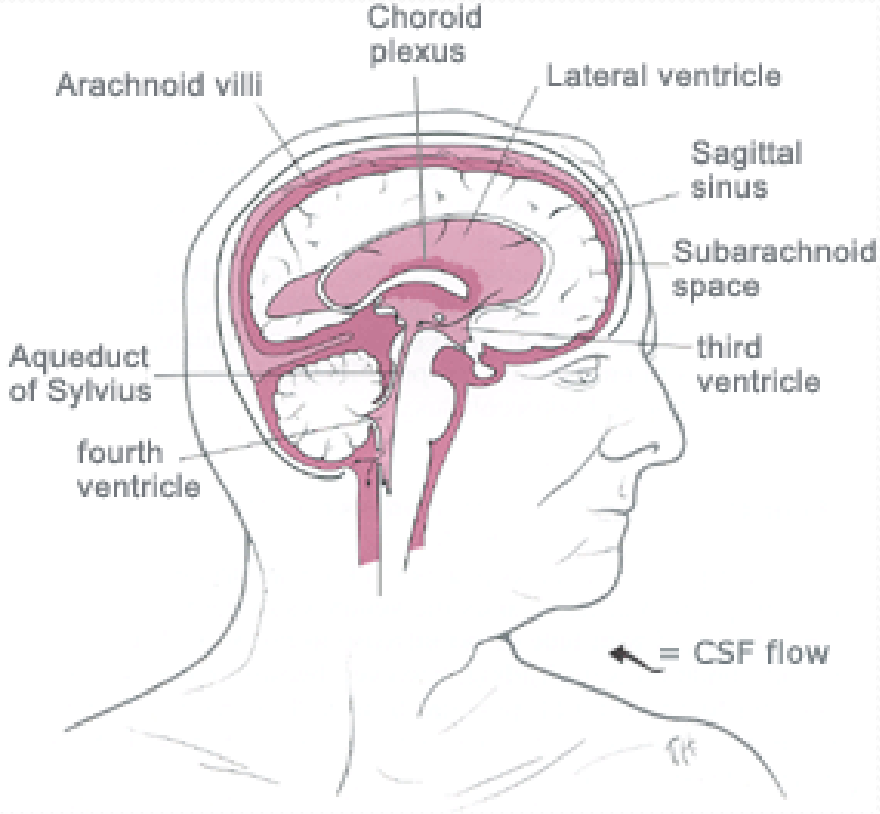






Overview of CSF circulation

- The CSF flows from the lateral ventricles downward to the foramina of Magendie and Luschka, to the perimedullary and perispinal subarachnoid spaces, and then upward to the basal cistern and finally to the superior and lateral surfaces of the cerebral hemispheres



NORMAL PRESSURE HYDROCEPHALUS

- First described by Adams,Fisher and Hakim in 1964
- Normal pressure hydrocephalus (NPH) is a clinical symptom complex characterized by abnormal gait, urinary incontinence, and dementia.

EPIDEMIOLOGY

- elderly individuals (age >65 y) the prevalence of NPH was 1.4%
- Incidence -1.4 per 1,00,000
- No race or gender prediliction
- more than 60% of patients with iNPH had cerebrovascular disease.
- In another similar study, more than 75% had Alzheimer disease pathology at the time of shunt surgery.

ETIOLOGY

- IDIOPATHIC – 50 %
- SECONDARY CAUSES – 50 %
- subarachnoid hemorrhage (23%),
- Resolved acute meningitis or a chronic meningitis(tubercular,fungal,syphilitic) (4.5%)
- and traumatic brain injury (12.5%)
- Congenital aqueductal stenosis
- Paget's disease of the base of skull
- Mucopolysaccharidosis of the meninges
- Achondroplasia

Pathophysiology

- ↓ CSF absorption at the arachnoid villi
- ↑ CSF pressure
- Force = Pressure X Area
- ↑ force against the brain than the same pressure in normal-sized ventricles
- Ventricular enlargement
- With further enlargement of the ventricles, CSF pressure returns to normal

PATHOPHYSIOLOGY

- Increased subarachnoid space volume does not accompany increased ventricular volume.
- Decreased global and regional cerebral blood flow in periventricular white mater,frontal grey mater,thalamus and in the basal ganglia
- Symptoms result from distortion of the central portion of the **corona radiata** by the distended ventricles. Interstitial edema of the white matter
- The periventricular white matter anatomically includes the sacral motor –abnormal gait and urinary symptoms
- **Dementia** results from distortion of the periventricular limbic system.

CLINICAL FEATURES

- CLASSIC TRIAD
- **GAIT DISTURBANCE** -is typically the *earliest feature noted and considered to be the most responsive to treatment*
- Apraxia of gait – no weakness or ataxia
- bradykinetic, broad based, magnetic, and shuffling
- Hydrocephalus Astasia abasia
- **URINARY SYMPTOMS** of NPH can present as urinary frequency, urgency, or frank incontinence.

- **DEMENTIA**
- **Usually mild**
- prominent memory loss and bradyphrenia.
- Frontal and subcortical deficits
- forgetfulness, decreased attention,
- Aphasia /agnosia – alternate diagnosis -Alzheimer

Proposed diagnostic criteria

Case of probable iNPH are

- Older than 40 years of age
- insidious (nonacute) progression of symptoms over a period of at least 3 months
- **CSF** opening pressures between 70 and 245 mm H_2O.
- **MRI or CT** must show ventricular enlargement as well as an Evan's index of at least 0.3 , periventricular signal changes, periventricular edema, or an aqueductal/fourth ventricular flow void

DIFFERENTIAL DIAGNOSIS

Disease	Common features with NPH	Features that are atypical of NPH
Cortical dementias		
Alzheimer's disease	dementia without gait impairment is very rare	No gait disturbance until dementia is at least moderately severe; focal cortical deficits
Fronto-temporal dementia		Personality change, psychiatric abnormalities: disinhibition, impulsiveness, irritability, emotional lability; aphasia; no motor disturbance; incontinence very rare
Subcortical dementias		
Lewy-body dementia	Gait impairment and dementia	Visual hallucinations, delusions, markedly fluctuating cognitive function
Parkinson's disease and vascular parkinsonism	Hypokinetic gait, tremor (40%) in NPH (unilateral symptoms in NPH always indicate some type of comorbidity [e.g., cerebrovascular disease, Alzheimer's disease])	Rest tremor, unilateral onset; speed of movement can be increased with the aid of external stimuli (this is not the case in NPH). The patient cannot simulate walking and bicycle-riding while supine; no broad-based gait with externally rotated feet; mildly reduced step height, markedly reduced arm swing, markedly stooped posture, autonomic dysfunction

Progressive supranuclear palsy	Frontal brain signs, impaired executive function, gait disturbance	Pseudobulbar palsy, supranuclear upward gaze paresis
Corticobasal degeneration		Rigor, asymmetrical symptoms, alien-limb phenomenon, apraxia, supra-nuclear upward gaze paresis, cortical sensory deficits, severe loss of postural control
AIDS-dementia complex	Psychomotor slowing, impairment of memory and concentration, gait impair-ment due to HIV myelopathy	Positive HIV serology
Age-related depression	Pseudodementia, neuropsy-chological test findings very	Depressive thought content because of frequently comorbid vascular

- Diagnosis of NPH less likely IF-
- ● Intracranial pressure above 25 cm H2O (this rules
- out iNPH, by definition) (Normal ICP is 5-15 cm of H2O)
- ● Age under 40 (iNPH unlikely)
- ● Asymmetrical or transient symptoms
- ● Cortical deficits, e.g., aphasia, apraxia, or paresis
- ● Progressive dementia without gait disturbance
- (even if the ventricles are enlarged)
- ● Lack of progression of symptoms

INVESTIGATIONS

- CSF – analysis –

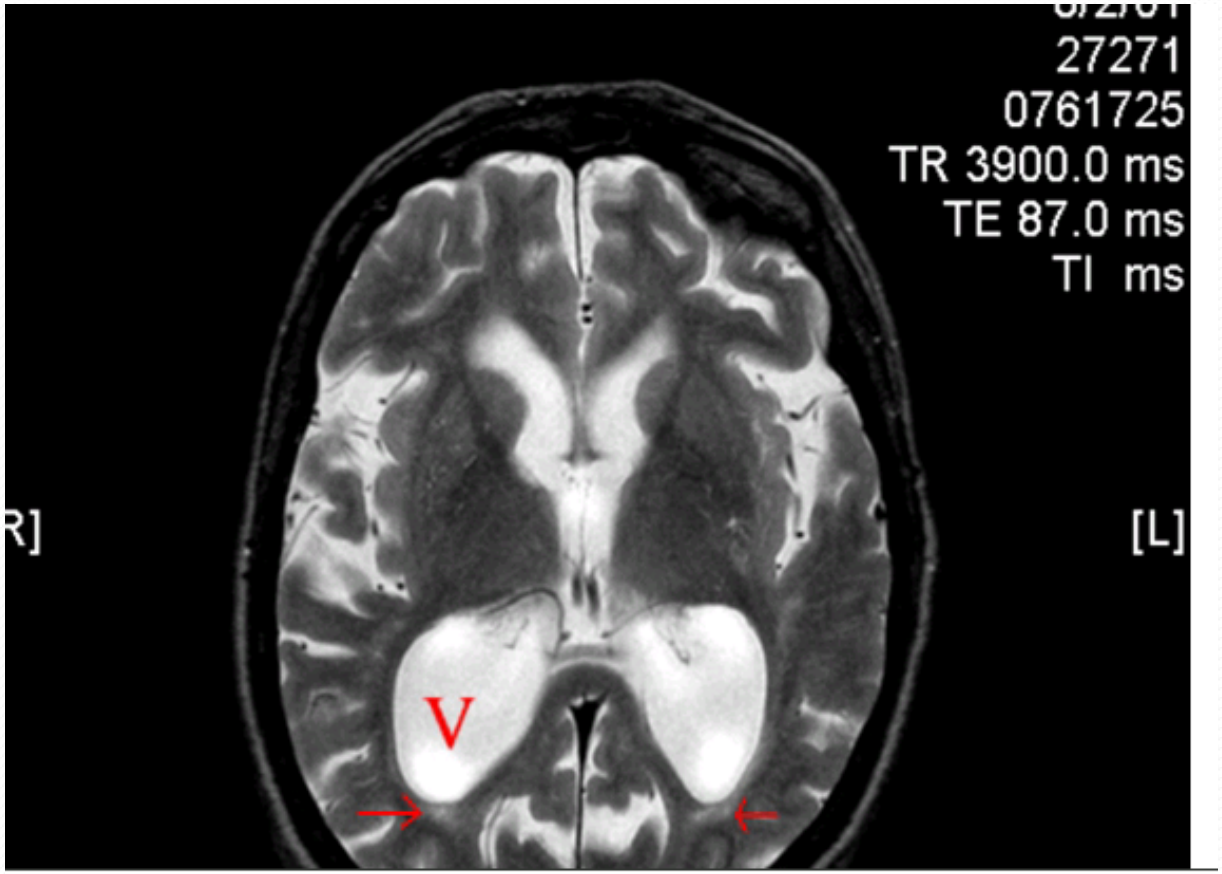
opening pressure – 150-200 mm H20 slightly higher than normal

- (CSF) protein Lipocalin-type prostaglandin D synthase (L-PGDS) – marker of Frontal lobe dysfunction in iNPH – decreased due to damage of arachnoid cells

- studies also showing that amyloid beta42 levels reduced in patients with NPH

CT SCAN

- Ventriculomegaly that is out of proportion to sulcal atrophy.
- Frontal and occipital periventricular hypoattenuating areas, represent transependymal CSF flow - **infrequent** and often may represent periventricular leukoencephalopathy
- corpus callosal thinning, -nonspecific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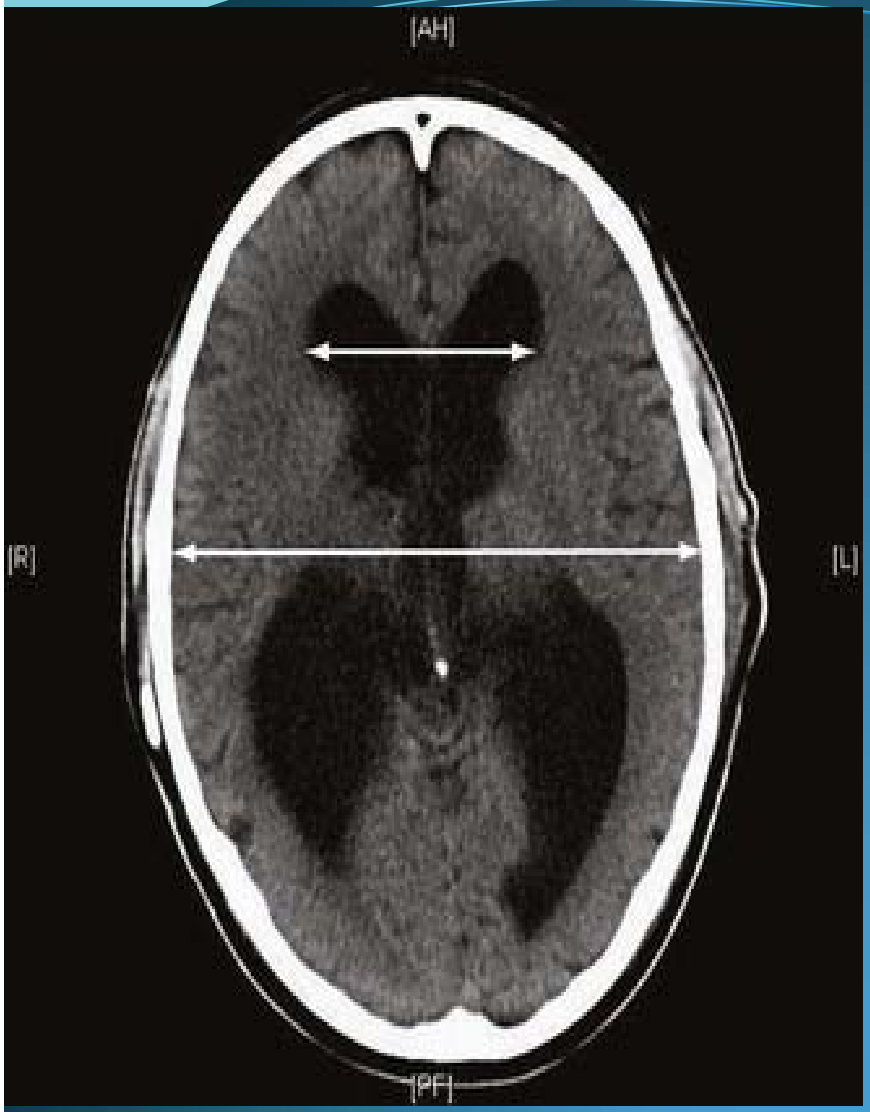
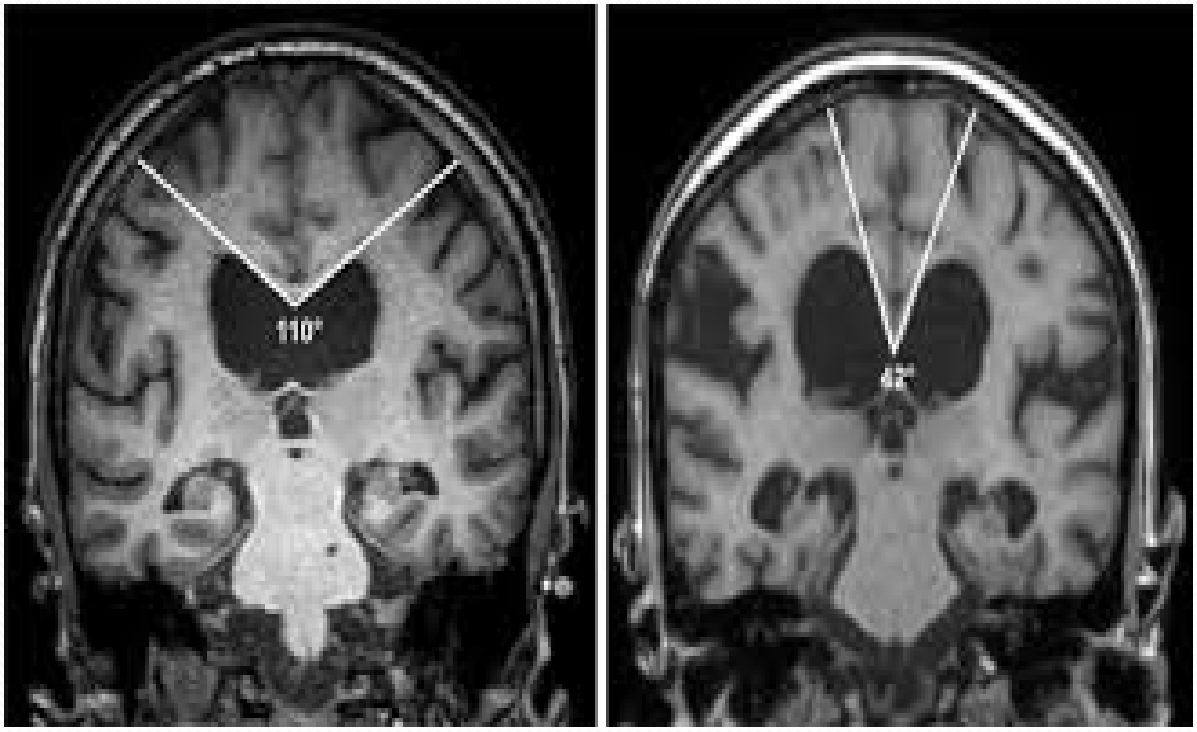


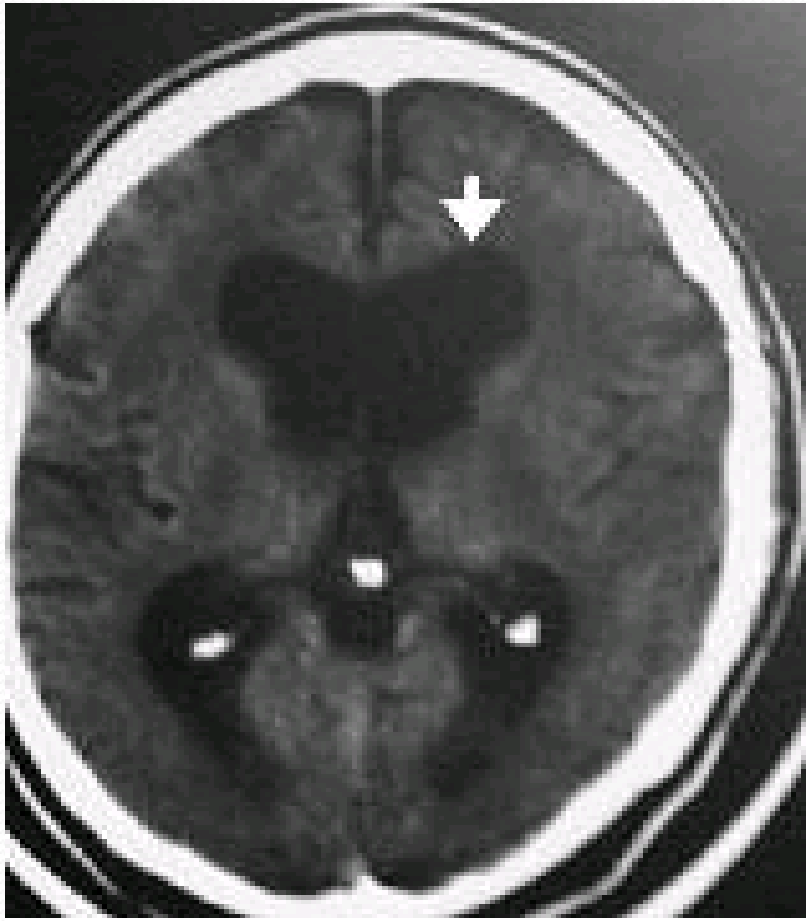
A frontal horn ratio (Evans' index), defined as the maximal frontal horn ventricular width divided by the transverse inner diameter of the skull, signifies ventriculomegaly if it is **0.3** or greater.

The **callosal angle** measured on MRI as a predictor of outcome in idiopathic normal-pressure hydrocephalus.



- Rounding of frontal horns

- clinical picture and ventriculosulcal disproportion on either CT or MRI scans, 50-70% of patients are likely to respond to a CSF-shunting procedure.



Magnetic Resonance Imaging

MRI provides additional physiologic information on NPH

- T2-weighted images, regions of moving CSF demonstrate no signal, instead of the increased signal observed in slow-moving CSF,
- **CSF flow studies**- jet of turbulent CSF flow may be observed distal to the aqueduct
- Cine phase-contrast MRI quantifies CSF flow in terms of stroke volume
- - significant corelation to shunt responsiveness

CSF TAP TEST

- Most prefer 45 -50 ml removal
- Csf pressures may be transiently elevated
- Improvement may be delayed and appear 1-2 days after
- Sensitivity of test – 62 % and 33 % specificity
- However it has been listed in guidelines of prognostic evaluation of NPH

EXTERNAL LUMBAR DRAINAGE

- greater impact on brain volume expansion
- 50ml/day drained over 3 days consecutively
- Complications -including headache, radiculopathy, and bacterial meningitis
- More sensitive than csf tap test
- sensitivity, specificity, and negative predictive value were 95%, 64%, and 78%, respectively.
- PPV 80 -100 %
- Requires hospitalisation specialised care

PRESURGICAL EVALUATION

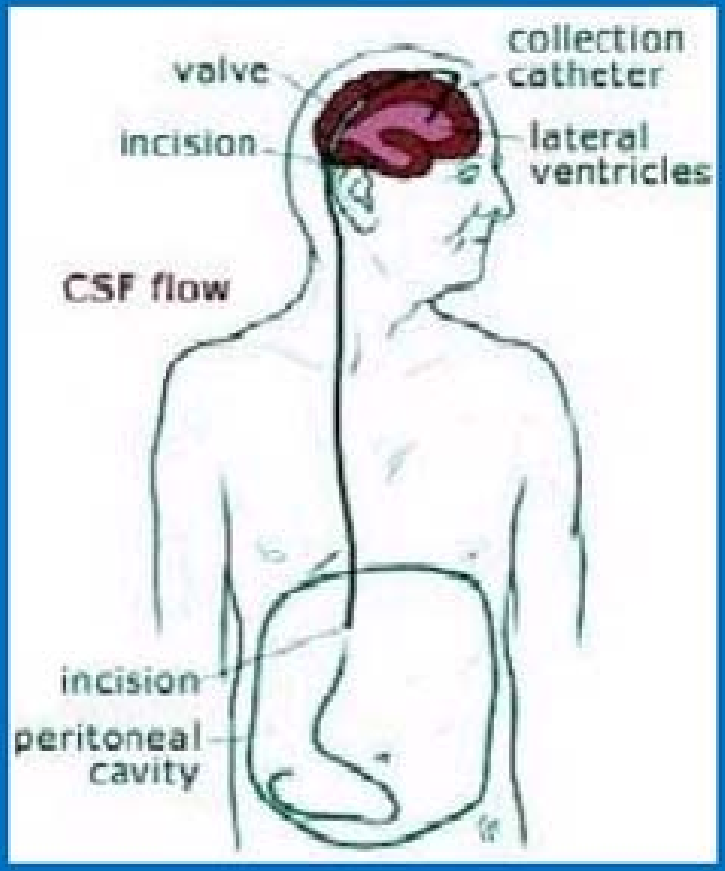
- Neuropsychological evaluation (eg, Folstein test or formal neuropsychological evaluation)- not validated
- Timed walking test.
- Videotaping the gait evaluation before and after the large volume lumbar puncture.- IS PREFERABLE
- Reduction in bladder hyperactivity also may be a sign of good outcome from shunting.

- Timed walking Test

MANAGEMENT

- No prospective, double blind, randomized, controlled clinical triaL
- Medical management – levodopa trial to rule out idiopthic parkinson disease
- No drug is known to work in NPH.Low dose Acetazolamide was used previously,but recent studies showed no efficacy.
- Surgical Management – mainstay
- Benefit expected from shunt surgery in probable case of NPH 50 %- 61 %

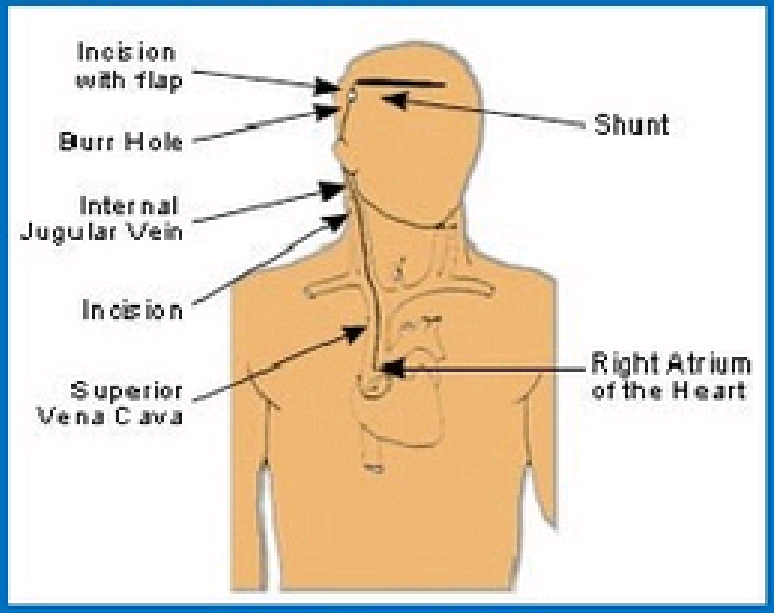
Surgical Management



- **Ventriculoperitoneal Shunt (VP shunt)**
  - Performed under general anaesthesia
  - Catheter placed within a ventricle, and another end at the peritoneal cavity

Venticuloatrial Shunt (VA Shunt)

- CSF is shunted from the cerebral ventricles into the right atrium of the heart.
- 2nd preferred choice if VP shunt is not possible
  - Eg. Infection of peritoneal cavity -> affects reabsorption rate of CSF



Complications Associated with Shunting

- Anesthesia-related risks (such as myocardial infarction)
- **Acute intracerebral hemorrhage** is the primary "procedure-related" risk
- Infection
- Seizures
- Shunt obstruction
- Subdural fluid collection and hematoma
- Overdrainage headaches
- Shunt underdrainage (failure to improve despite a patent shunt).

- Mean rate of complications was 38 percent
- Additional surgery required in 22 %
- The decision for surgery should be individualised

- Ileus
  - Slow gastric and bowel movement post operation and may feel nausea
- Infection
  - Most common organisms are *S. epidermidis and S. aureus*
- Obstruction
  - Most often due to the head tip is obstructed with cells, choroid plexus, or debris.

Newer advances

- adjustable shunt valves – adjusts the opening pressure
- the introduction of gravity-controlled valves - low valve opening pressure when the patient is lying down.
- G valves lower the risk of overdrainage by 90%

Response to Surgical Intervention

- Many, if not most, INPH patients have comorbid brain conditions.
- Periventricular white matter ischemia is commonly seen in INPH patients. Patients with severe cerebrovascular disease do not respond as well to shunting but may still derive some benefit from the procedure.
- The neurological decline sometimes seen despite shunt placement in INPH may be related to the progression of comorbid conditions
- Even temporary improvements ranging from 1 to 3 years may make a substantial difference in quality of life.

FOLLOW UP

- Routine follow up 2 to 3 times per year
- Earlier if shunt inection/failure
- Bedside clinical examination follow up CTScan within few weeks
- D Dimer ,CRP in case of ventriculoatrial shunts for subclinical septicemia and thromboembolism

Whether to shunt or Not?

- High CSF pressure should prompt investigation for a secondary cause of NPH
- Response to a 40-mL to 50-mL (high-volume) lumbar tap suggests a potential benefit to shunting
- An ELD may be used to evaluate those who do not respond to a high-volume tap
- There is no substantial predictive value to MRI CSF flow studies
- IF multi-infarct or Alzheimer's disease dementia ??

THANK YOU