hydrocephalus

Fb/Nurse-Info

Definition
HYDROCEPHALUS

It is defined as a disproportionate increase in the amount of CSF within the cranium, usually in association with a rise in ICP.

(Ventricular enlargement with excessive CSF) And it should be differentiated from hydrocephalus *ex vacuo* (due to brain atrophy)

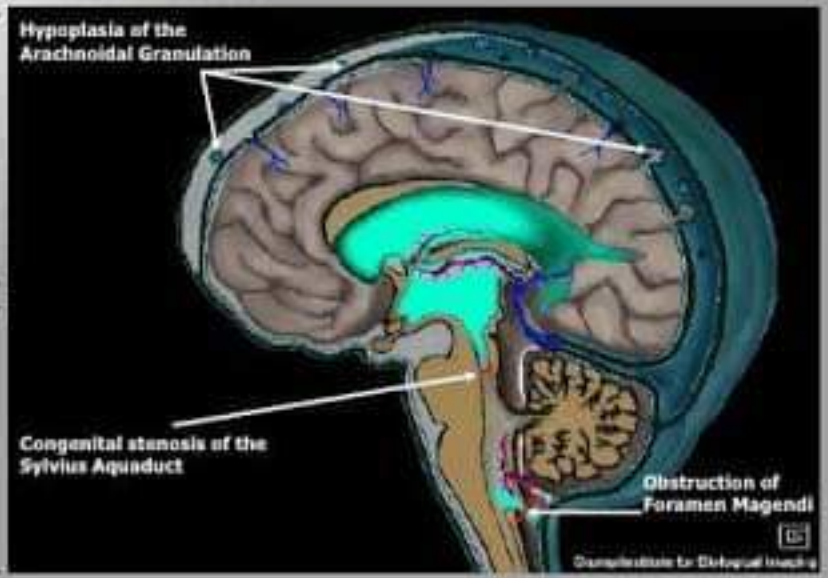
TWO TYPES OF HYDROCEPHALUS

Communicating

- Communicating (non-obstructive) hydrocephalus is the situation where there is communication between the ventricular system and the subarachnoid space
- The most common cause of this group is post-infective and post-haemorrhagic hydrocephalus

Non-communicating

- Non-communicating or obstructive hydrocephalus is where there is no communication between the ventricular system and the subarachnoid space
- The most common cause of this category is aqueduct blockage



CSF

What causes hydrocephalus?

- The causes of hydrocephalus are still not well understood. Hydrocephalus may result from inherited genetic abnormalities (such as the genetic defect that causes aqueductal stenosis) or developmental disorders (such as those associated with neural tube defects including spina bifida and encephalocele). Other possible causes include complications of premature birth such as intraventricular hemorrhage, diseases such as meningitis, tumors, traumatic head injury, or subarachnoid hemorrhage, which block the exit of CSF from the ventricles to the cisterns or eliminate the passageway for CSF into the cisterns.

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#Typeses of hydrocephalus

- Hydrocephalus may be congenital or acquired. Congenital hydrocephalus is present at birth and may be caused by either events or influences that occur during fetal development, or genetic abnormalities. Acquired hydrocephalus develops at the time of birth or at some point afterward. This type of hydrocephalus can affect individuals of all ages and may be caused by injury or disease.

Introduction

In the earlier last million years of the cenezoic period, human-like people lived and roamed earth. They evolved and evolved until they finally became just like us! But while they still were “cave men”, they used something that every single modern day person needs to count on. Tools. Even millions of years ago, these creatures used tools to help them survive, whether its poking a stick in an ants nest or making weapons to hunt holy mammoths.

Etiology

- Congenital …..?
  - aqueduct stenosis
  - Chiari malformations
  - Dandy-Walker malformation
- Acquired
  - infection (post-meningitis)
  - post – hemorrhagic(SAH,IVH)
  - secondary to masses(non- neoplastic like vascular malformation, tumors like posterior foss tumors, pituitary and colloid cyst)

Post-Meningitic Hydrocephalus

- Hydrocephalus is a significant cause of childhood mortality and morbidity
- Bacterial meningitis is the largest acquired cause of non-tumoural hydrocephalus
- Little published population based data on the incidence of hydrocephalus after meningitis
- Single hospital series suggest hydrocephalus complicates 1.7- 2.8% of episodes of bacterial meningitis in children *(Jadavji T, 1986, Thomas DJ, 1989)*

Post meningitic hydrocephalus

- 513 children with bacterial meningitis were notified to CDSC
- 129 cases of severe bacterial meningitis were admitted to Regional PICU
- 11 children (6 boys, 5 girls) underwent shunt surgery for post-meningitic hydrocephalus at RNSC
- Incidence of hydrocephalus requiring VP shunt 11/513 (2.1%) cases of meningitis, 11/129 (8.5%) of cases requiring PICU

Prevalance: 1-1.5%
Incidence: 0.3-3.5%
Upto 20% after SAH
1% after meningitis

CSF:

TOTAL VOLUME :
NEONATES : 50ml
ADULT : 150ml
{50%- CRANIAL CAVITY
50%- SPINAL CAVITY}

RATE :
NEWBORN -20ml/day
ADULT- 500ml/day

PRESSURE:
ADULT – 90-180mm H20
NEWBORN- 10-100mm H2O

TURNOVER – 3TIMES/DAY

FOLLOW UP:

- HC MONITORING
- SIGNS OF INFECTION/BLOCK
- ANTIBIOTIC PROPHYLAXIS
- RE-EVALUATION

PROGNOSIS:

depends on primary cause
Large IVH- permanent hydrocephalus
NPH- responds to shunt
gait and incontinence respond to shunting
but dementia responds less frequently.

PARINAUD SYNDROME

CLINICAL FEATURES :

Signs :

Macrocephaly
Dysjunction of sutures
Dilated scalp veins
Skin over scalp thin and shiny
MACEWEN SIGN
Tense/bulging fontanelle
SETTING-SUN SIGN
Pyramidal signs
Lateral rectus palsy
Papilledema
Bradycardia
Altered respiration
Cushing triad

Symptoms :

Poor feeding
High pitched cry
Irritability
Reduced activity
Vomiting

Headache
Bladder incontinence
Blurred vision
Drowsiness
Horizontal diplopia

Congenital Hydrocephalus

- Congenital hydrocephalus is present at birth and may be caused by environmental influences during fetal development or by genetic factors.
- Causes are:
  - Aqueduct Stenosis
  - Colloid Cyst

Congenital Hydrocephalus

- Incidence: 1/1000-2/1000
- Etiology
  - "Communicating" hydrocephalus
  - Meningomyelocele with Chiari II anomaly
  - Aqueductal stenosis
  - Dandy-Walker malformation
  - Other
- 15% have aqueductal stenosis
  - Sibling risk is 10% when male affected
- Sibling risk is 4% for uncomplicated types

Pathophysiology of hydrocephalus

- Acute hydrocephalus

-in animals…

- In humans :

1. ICH
2. Acute decompensation of tumors bordering the ventricular system
3. Gunshot
4. SAH
5. Head injuries
6. Sudden shunt obstruction

If the CSF obstruction is complete , ventricular enlargement may reach significant proportions in 3 to 6 hr.

3. Clinical

- Clinical – Gait disturbance ± Cognition impairment ± Urinary symptoms
- With respect to **gait/balance**, at least two of the following should be present and not be entirely attributable to other conditions
  a) Decreased step height
  b) Decreased step length
  c) Decreased cadence (speed of walking)
  d) Increased trunk sway during walking
  e) Widened standing base
  f) Toes turned outward on walking
  g) Retropulsion (spontaneous or provoked)
  h) En bloc turning (turning requiring three or more steps for 180 degrees)
  i) Impaired walking balance, as evidenced by two or more corrections out of eight steps on tandem gait testing

Treatment:

- Hydrocephalus is most often treated by surgically inserting a shunt system. This system diverts the flow of CSF from the CNS to another area of the body where it can be absorbed as part of the normal circulatory process.
- A shunt is a flexible but sturdy plastic tube. A shunt system consists of the shunt, a catheter, and a valve. One end of the catheter is placed within a ventricle inside the brain or in the CSF outside the spinal cord. The other end of the catheter is commonly placed within the abdominal cavity, but may also be placed at other sites in the body such as a chamber of the heart or areas around the lung where the CSF can drain and be absorbed. A valve located along the catheter maintains one-way flow and regulates the rate of CSF flow.

HYDROCEPHALUS : SIGNS & SYMPTOMS

- Eyes that appear to gaze downward;
- Irritability;
- Seizures;
- Separated sutures;
- Sleepiness;
- Vomiting.
- Brief, shrill, high-pitched cry;
- Changes in personality, memory, or the ability to reason or think;
- Changes in facial appearance and eye spacing;
- Loss of bladder control (urinary incontinence);
- Loss of coordination and trouble walking;
- Muscle spasticity (spasm);

Symptoms

Spina Bifida

- Paralysis and muscle weakness.
- Difficulty reading and solving problem.
- Bowel incontinence and urinary incontinence.

Hydrocephalus

- Usually large head.
- Seizures.
- Eyes fixed downward.
- Developmental delay.
- Headache followed by vomiting.
- Memory loss.
- Drowsiness.
- Changes in personality.

Causes of communicating hydrocephalus

- Overproduction of CSF
- Blockage of CSF circulation
- Blockage of CSF resorption
- Hydrocephalus ex-vacuo
- Normal pressure hydrocephalus

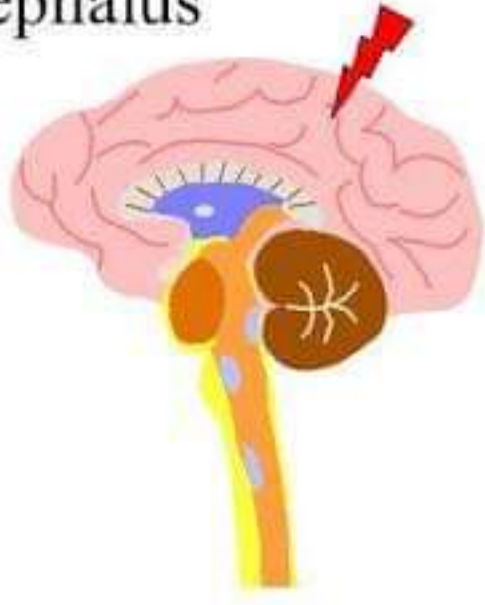

Table 2. Differentiating Between Hydrocephalus and Diseases With Similar Presentation

Disease State	Symptoms Shared With Hydrocephalus	Symptoms Not Shared With Hydrocephalus
Alzheimer's disease	Decreased memory function Personality changes Incontinence Cognitive impairment Possible gait disturbances	Social withdrawal Usually begins in one's 60s Aphasia and apraxia Difficulty sleeping
Parkinson's disease	Gait disturbances Confusion Cognitive impairment	Gait disturbances responsive to levodopa Cogwheel rigidity Sialorrhea
Creutzfeldt-Jakob disease	Confusion Gait disturbances Decreased coordination Muscle spasms Extreme sleepiness Personality changes	No loss of bladder control Usually begins in one's late 50s Hallucinations
Vascular dementia	Confusion Cognitive impairment Incontinence	Emotional instability Possible signs of stroke on MRI Difficulty sleeping

Source: Reference 8.

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- **Communicating hydrocephalus** – Most common complication of TBM and is caused by blockage of the basal cisterns by inflammatory exudates.

- In some cases, the hydrocephalus may be noncommunicating, resulting from obstruction due to tuberculoma or rarely tubercular abscess.

- **Ischemic infarcts** are also common, seen in 20-40% cases, mostly within basal ganglia and internal capsule regions, resulting from vascular compression and occlusion of small perforating vessels, particularly Lenticulostriate and Thalamoperforating arteries. (**Necrotizing Arteritis**).

Ventriculitis (non-communicating)

Ventriculitis is a disease causing inflammation and or infection of the ventricles. It is most common in infants and is

CAUSES OF HYDROCEPHALUS

- OBSTRUCTIVE HYDROCEPHALUS

➢ Congenital

- Aqueductal stenosis
- Arnold chiari syndrome
- Dandy-walker syndrome

➢ Acquired

- Aqueductal gliosis-meningitis,bleeding,mumps encephalitis
- Posterior fossa tumors-medulloblastoma

CAUSES OF HYDROCEPHALUS

- COMMUNICATING HYDROCEPHALUS
  - Increased Production
    - Tumors in choroid plexus
  - Decreased Absorption
    - Congenital-TORCH
    - Acquired-Meningitis,leukemia

Communicating Hydrocephalus

- Causes are due to:
  - Post-hemorrhage
  - Bacterial Meningitis
  - Malignant Meningitis
  - Increased Venous Pressure

Non-Communicating Hydrocephalus

- Non-communicating hydrocephalus, also called "obstructive" hydrocephalus, occurs when the flow of CSF is blocked along one or more of the narrow pathways connecting the ventricles.
- Causes include:
  - Congenital
  - Acquired

Physiology and circulation of CSF

- *The **normal volume** of circulating CSF is about 150 ml.*
- ***The daily production** of the CSF is about 450 ml, so the CSF volume is replaced approximately three times daily.*
- *CSF is produced by an **active process** independent of inracranial pressure (ICP).*
- *80% of CSF is produced by the **choroid plexus**, and the rest is from the parenchyma*

Aetiologty of Hydrocephalus

- *In patients with hydrocephalus, an imbalance has occurred between the normal physiological production of CSF and its absorption.*
- *This imbalance can be as a result of overproduction of CSF, an obstruction, or impaired absorption.*

Treatment

Surgical therapy

- Principle of surgical intervention :

Relief increased ICP by diversion the exessive CSF from ventricular system into an absorptive surface out side the brain such as pleura or peritoneum or into the atria of the heart…….this is called **shunt operation**

- The most indication for shunt operation is progressive hydrocephalus .

COMPLICATIONS

1. Infection is the most feared complication
2. Subdural hematomas .
3. Shunt failure is mostly due to suboptimal proximal catheter placement .
4. Overdrainage is more common in lumboperitoneal shunts .
5. Slit ventricle syndrome .

Treatment

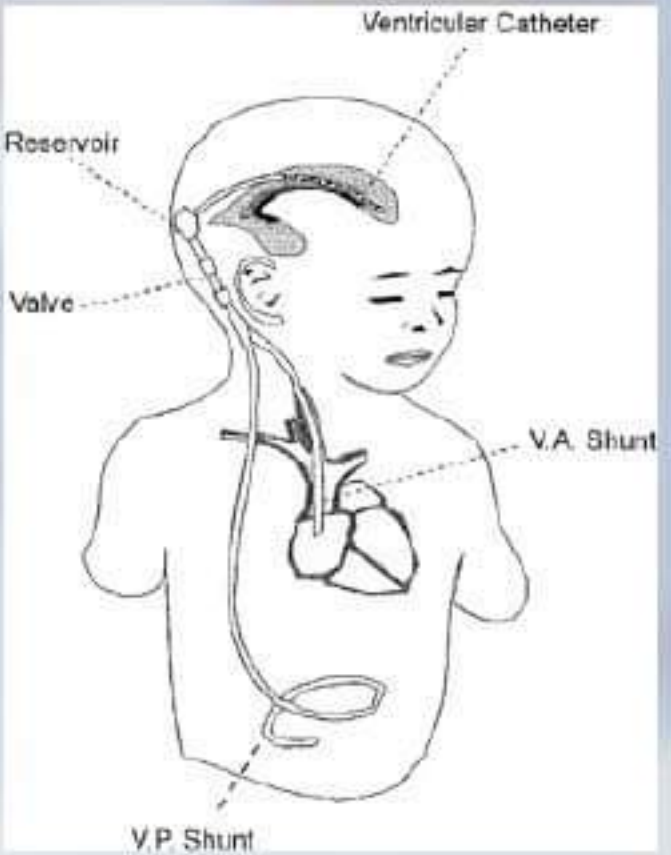
- Types of shunts :

1. Third ventriculostomy
2. Ventriculoperitoneal shunting (the common procedure) .
3. Ventriculoatrial shunting .
4. Ventriculopleural shunting .
5. Torkildsen shunts or internal shunts .
6. Lumboperitoneal shunts .

The 4 parts of a shunt

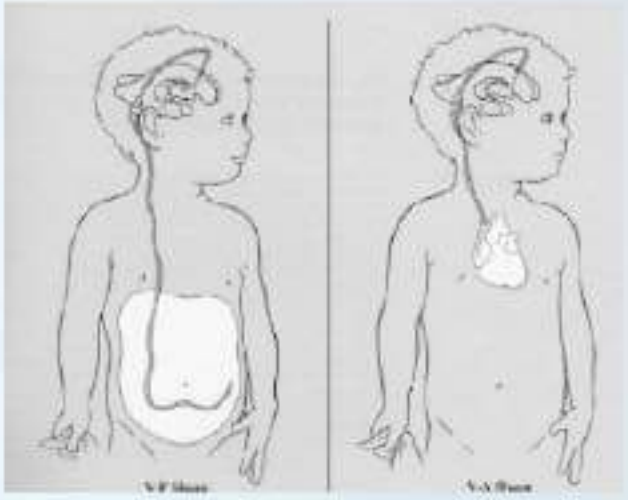
- **Ventricular *(Upper)* Catheter**- This is the top-most part of the shunt. It is a small, narrow tube that is inserted into the ventricle (a small opening or pouch) inside the brain that contains the cerebrospinal fluid (CSF).

- **Reservoir**-This is where the excess CSF is collected until it drains into the bottom portion of the shunt. The reservoir also lets the doctor remove samples of CSF for testing, and to inject fluid into the shunt to test for flow and to make sure the shunt is working properly.



Types of shunts

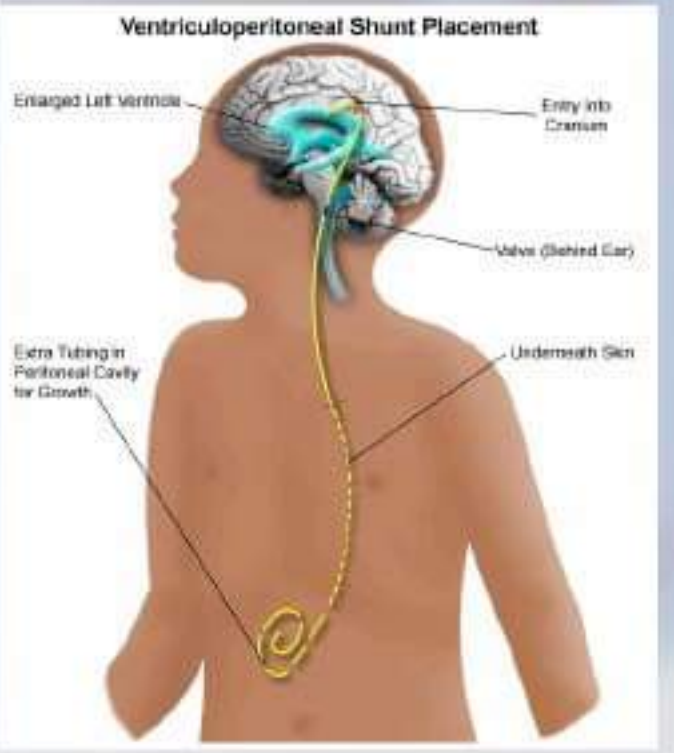
- Shunts are named according to where they are inserted in the brain and where they are inserted to let the excess CSF drain out.



- A ventriculo-peritoneal (VP) shunt drains into the abdomen or peritoneum (belly). Most shunts, including Sean's, are VP shunts.
- A ventriculo-pleural shunt drains into the space surrounding the lung.
- A ventriculo-atrial (VA) shunt drains into the atria of the heart.

What is a shunt?

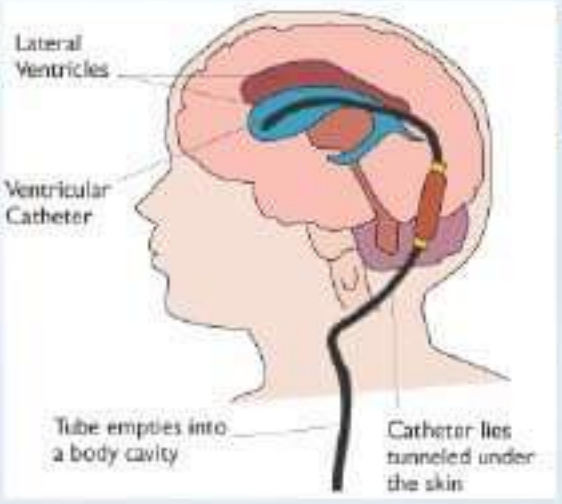
- A shunt is a piece of soft, flexible plastic tubing that is about 1/8-inch (3mm) in diameter.
- It allows excess cerebrospinal fluid (CSF) that has built-up inside the skull to drain out into another part of the body, such as the heart or abdomen.
- To drain excess CSF, shunts are inserted into an opening or pouch inside the brain called a ventricle, just above where the blockage is that is preventing the CSF from flowing properly.



How a shunt works

All shunts perform two functions.

- **They allow CSF to flow in only one direction, to where it is meant to drain.**
- **They all have valves, which regulate the amount of pressure inside the skull.**
  - **When the pressure inside the skull becomes too great the valve opens, lowering the pressure by allowing excess CSF to drain out.**



Signs that a shunt needs replacing:

- Loss of appetite
- Nausea and vomiting
- Abdominal pain or cramps
- Changes in mood, including being irritable
- Frequent or persistent headaches with increased severity
- Difficulty walking
- Numbness on one side of the body
- Muscle tension
- Sudden, constant, or extreme tiredness
- Difficulty thinking clearly or remembering
- Difficulty seeing or speaking
- Persistent fever
- Redness, swelling, or tenderness where the shunt is under the skin
- Coma
- Difficulty breathing
- Abnormal heart rate

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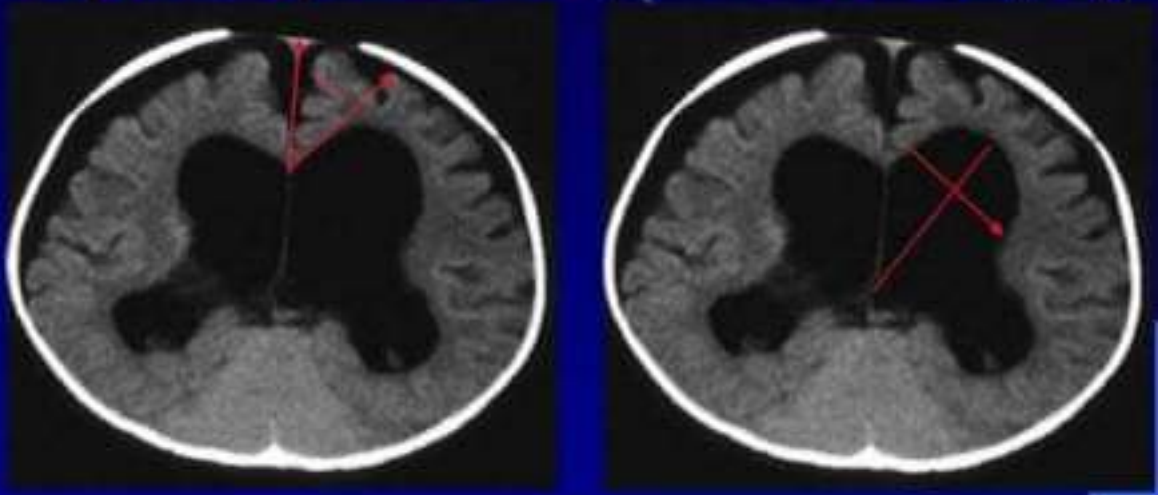
Investigation

- Ultrasonography to visualize the ventricular system(when the anterior fontanelle is patent). Also for antenatal diagnosis
- CT and /or MRI of the head; it is important to exclude any abnormal masses and to study the size and the shape of the ventricles, and some time needs contrast study.
- LP in cases of communicating hydrocephalus for both diagnostic and therapeutic…..(NPH)

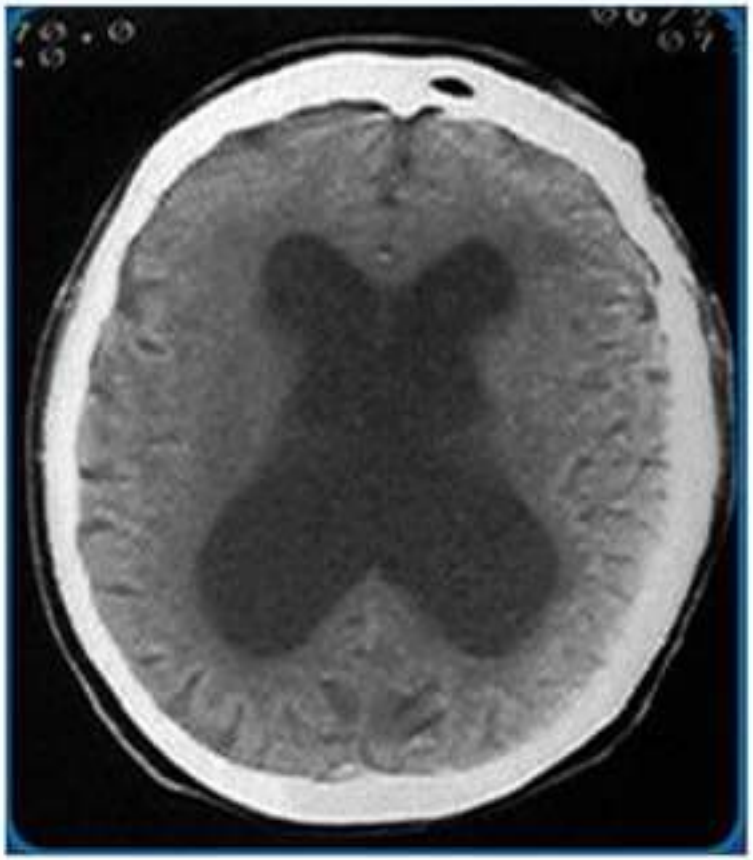
Hydrocephalus v. Cerebral Atrophy



Ventricular Angle (left) tends to be smaller in hydrocephalus (shown above) than in atrophy. Frontal horn radius (right) tends to be larger in hydrocephalus than in atrophy.

Brain Imaging

Normal Brain



NPH Brain