Hydrocephalus

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What is hydrocephalus?

- ▶ Greek words
- "hydro" meaning water and "cephalus" meaning head
- excessive accumulation of fluid in the brain
- "water" is actually cerebrospinal fluid (CSF)--a clear fluid that surrounds the brain and spinal cord

Why it is important to know

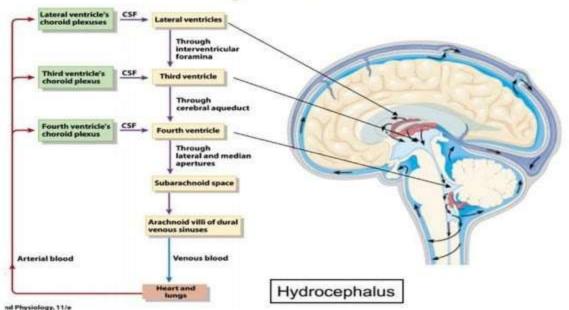
- The excessive accumulation of CSF results in an abnormal widening of spaces in the brain called ventricles.
- This widening creates potentially harmful pressure on the tissues of the brain.

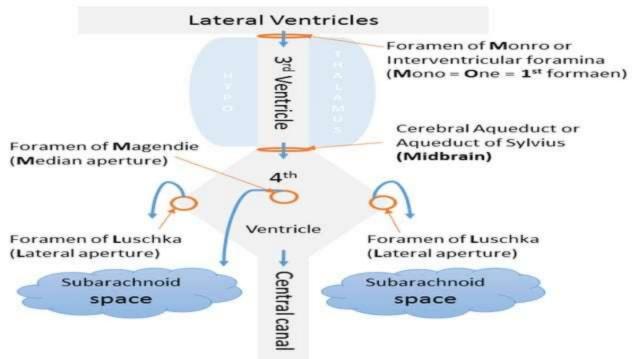
Normal Hydrocephalic

ventricular system

The ventricular system is made up of four ventricles connected by narrow passages.. Normally, CSF flows through the ventricles, exits into cisterns (closed spaces that serve as reservoirs) at the base of the brain, bathes the surfaces of the brain and spinal cord, and then reabsorbs into the bloodstream.

Pathway of CSF flow





Function of CSF

- Three important life-sustaining functions:
- 1) to keep the brain tissue buoyant, acting as a cushion or "shock absorber"
- 2) to act as the vehicle for delivering nutrients to the brain and removing waste
- 3) to flow between the cranium and spine and compensate for changes in intracranial blood volume.

Pathogenesis

- Normal volume of circulating CSF is around 150ml Daily production is about 500ml/day, ~20ml/hr
- The balance between <u>production</u> and <u>absorption</u> of CSF is critically important.
- Because CSF is made continuously, medical conditions that block its normal flow or absorption will result in an overaccumulation of CSF. The resulting pressure of the fluid against brain tissue is what causes hydrocephalus.

Types of hydrocephalus

Congenital or Acquired.

Congenital hydrocephalus is present at birth and may be caused by either events or influences that occur during foetal 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

- communicating or non-communicating
- Communicating hydrocephalus occurs when the flow of CSF is blocked after it exits the ventricles. This form is called communicating because the CSF can still flow between the ventricles, which remain open.

Non-communicating hydrocephalus - also called "obstructive" hydrocephalus - occurs when the flow of CSF is blocked along one or more of the narrow passages connecting the ventricles. One of the most common causes of hydrocephalus is "aqueductal stenosis." In this case, hydrocephalus results from a narrowing of the aqueduct of Sylvius, a small passage between the third and fourth ventricles in the middle of the brain.

Other types

- Hydrocephalus ex-vacuo and normal pressure hydrocephalus.
- <u>Hydrocephalus ex-vacuo</u> occurs when stroke or traumatic injury cause damage to the brain. In these cases, brain tissue may actually shrink and space is filled with csf, which may appers like hydrocephalus but its dilatation of ventricle system without pressure (pseudo hydrocephalus)
- Normal pressure hydrocephalus can happen to people at any age, but it is most common among the elderly. It may result from a subarachnoid hemorrhage, head trauma, infection, tumor, or complications of surgery. However, many people develop normal pressure hydrocephalus even when none of these factors are present for reasons that are unknown.

► <u>Acute</u>

Develops within days or few weeks - Manifests with rapid progression of symptoms - Requires early attention and treatment hydrocephalus caused by tumor.

► Chronic

- Over months (or even years) Subtle signs of memory impairment, walking difficulty, urinary incontinence –
- A classic example is NPH Chronic hydrocephalus can present acutely because of changes in the pathophysiology of the CSF absorption or flow.

Causes

- not well understood.
- Congenital
 - Present at birth or few weeks/months after birth (1-2/1000 live births) Aqueduct stenosis Chiari malformations Dandy-Walker malformation
 - inherited genetic abnormalities Aqueductal stenosis
 - Developmental disorders neural tube defects including spina bifida and encephalocele
 - Complications of premature birth Intraventricular hemorrhage,
- Acquired
 - Infection (post-meningitis)
 - ▶ Post traumatic hemorrhagic (SAH,IVH)
 - ► Tumors
 - Subarachnoid haemorrhage which block the exit of CSF from the ventricles to the cisterns or eliminate the passageway for CSF into the cisterns.

Etiology	%
Intraventricular hemorrhage	24
Myelomeningocele	21
Brain tumors	9
Aqueduct stenosis	7
CSF infection	5
Head injury	2
Others	11
Unknown	12

Symptoms of hydrocephalus

Symptoms of hydrocephalus vary with age, disease progression, and individual differences in tolerance to the condition.

For example, an infants ability to compensate for increased CSF pressure and enlargement of the ventricles differs from an adults. The infant skull can expand to accommodate the build up of CSF because the sutures (the fibrous joints that connect the bones of the skull) and fontanelle that have not yet closed.

In infancy

- The most obvious indication of hydrocephalus is often a rapid increase in head circumference or an unusually large head size.
- Other symptoms
 - vomiting, sleepiness, irritability, downward deviation of the eyes "sun set eyes", seizures.
 - Fontanel full, bulging and wide Thin and glistening scalp with enlargement and
 - engorgement of scalp veins
 - Macewen's sign (cracked pot sound on head percussion)
 - Sixth nerve (abducens) palsy Setting sun sign upward gaze palsy
 - Hyperactive reflexes.
 - Irregular respiration with apneic spells.
 - Separation of cranial sutures (sutures diastasis)



In Older children and adults

- may experience different symptoms because their skulls cannot expand to accommodate the build up of CSF.
- Symptoms may include
 - headache followed by vomiting.
 - nausea.
 - papilledema (swelling of the optic disk which is part of the optic nerve).
 - blurred or double vision.
 - sunsetting of the eyes.
 - problems with balance,
 - poor coordination, gait disturbance, urinary incontinence,
 - slowing or loss of developmental progress, lethargy, drowsiness, irritability, or other changes in personality or cognition including memory loss,

Normal pressure hydrocephalus

- Symptoms of normal pressure hydrocephalus include, problems with walking, impaired bladder control leading to urinary frequency and/or incontinence, and progressive mental impairment and dementia.
- Classic triad of clinical findings (known as the Adams triad or Hakim's triad)
 - urinary incontinence,
 - gait disturbance, and
 - dementia
- commonly referred to as "wet, wacky and wobbly"

Diagnostic modalities

- History and general examinations
- Ultrasonography to visualize the ventricular system
 (when the anterior fontanelle is patent)
- Computed tomography (CT).
- Magnetic resonance imaging (MRI).
- Pressure-monitoring techniques.
- Spinal tap or lumbar catheter.
- Intracranial pressure monitoring.
- neuropsychological tests, to help them accurately diagnose normal pressure hydrocephalus and rule out any other conditions.

Obstructive or non communicating hydrocephalus



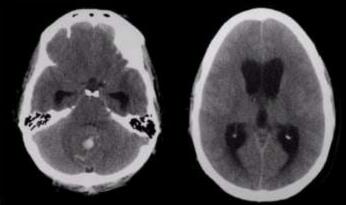
Communicating hydrocephalus



CT scan of colloid cyst (large white arrow) of the third ventricle with associated non-communicating hydrocephalus



Post fossa tumor



Treatment

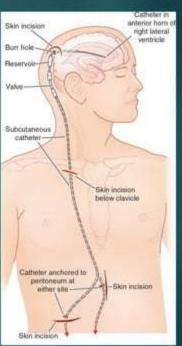
- The ultimate goal is to reverse the neurologic damage caused by the raised ICP
- Medical treatment not proved to be useful but Used as a temporary measure and in conjunction with surgical management.
 - Acetazolamide Commonly used reduce CSF production. However, benefits are minimal and high doses of the drug, which cause metabolic acidosis, are required to achieve the effect.
 - ▶ Dose 50mg/kg/day diminishes CSF production
 - Oral glycerol has also been used for the similar purpose

Treatment

- Hydrocephalus is most often treated by surgically
- Removal of the obstruction tumor, cyst excision.
- <u>CSF Diversion</u> 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.
 - Ventriculo-peritoneal (VP) most common
 - Ventriculo-atrial (VA) rarely done
 - Ventriculo pleural shunt
 - Lumbo-peritonial shunt.
 - External drainage temporary
- 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.

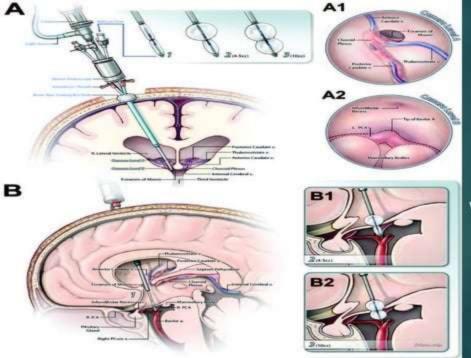
Shunt system





Third ventriculostomy

- A limited number of individuals can be treated with an alternative procedure called third ventriculostomy. In this procedure, a neuroendoscope — a small camera that uses fiber optic technology to visualize small and difficult to reach surgical areas
- Once the scope is guided into position, a small tool makes a tiny hole in the floor of the third ventricle, which allows the CSF to bypass the obstruction and flow toward the site of resorption around the surface of the brain.



Third Ventriculostomy

Complications of a shunt system

- Shunt systems are not perfect devices.
- Complications may include mechanical failure, infections, obstructions, and the need to lengthen or replace the catheter.
- Generally, shunt systems require monitoring and regular medical follow up. When complications occur, the shunt system usually requires some type of revision.
- Some complications can lead to other problems such as overdraining or underdraining.
- Overdraining occurs when the shunt allows CSF to drain from the ventricles more quickly than it is produced. Overdraining can cause the ventricles to collapse, tearing blood vessels and causing headache, hemorrhage (subdural hematoma), or slit-like ventricles (slit ventricle syndrome).

- <u>Underdraining</u> occurs when CSF is not removed quickly enough and the symptoms of hydrocephalus recur.
- In addition to the common symptoms of hydrocephalus, infections from a shunt may also produce symptoms such as a low-grade fever, soreness of the neck or shoulder muscles, and redness or tenderness along the shunt tract. When there is reason to suspect that a shunt system is not functioning properly (for example, if the symptoms of hydrocephalus return.

Prognosis

- The prognosis for individuals diagnosed with hydrocephalus is difficult to predict, although there is some correlation between the specific cause of the hydrocephalus and the outcome.
- Prognosis is further complicated by the presence of associated disorders, the timeliness of diagnosis, and the success of treatment.

Take home message

- The success of treatment varies from person to person, some people recover almost completely after treatment and have a good quality of life.
- Early diagnosis and treatment improves the chance of a good recovery.
- Dilated ventricles without pressure symptoms is ventriculomegaly not hydrocephalus.

Thank you..

Have a nice day.