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Hydrocephalus



What is the Definition of hydrocephalus?

Hydrocephalus is an abnormal buildup of cerebrospinal fluid (CSF) in the ventricles (cavities) inside the brain. This abnormal amount of CSF causes the ventricles to be larger than normal, putting an extra and abnormal amount of pressure inside the rigid skull. Hydrocephalus may present at or after birth (congenital) or later because of an infection, bleeding, or a mass inside the brain that can develop at any age (acquired). Acute hydrocephalus is an emergency that requires an immediate surgical intervention to prevent elevated intra-cranial pressure (ICP) inside the rigid skull, hence preventing further consequences.



An animated picture showing normal ventricles on the right, and dilated ventricles on the left (Hydrocephalus)

What is cerebral spinal fluid?

CSF is the clear fluid that circulate inside the central nervous system that works to protect the nervous system and works as a cushion to the brain and spine. CSF flows from the ventricles and circulate around the brain and spinal cord before getting reabsorbed into the blood. The body naturally produces a specific amount of CSF every day and absorbs it in the same amount to keep the pressure at the same level. Excess production of CSF can prevent the brain from functioning properly, and eventually may lead to brain damage or death in some cases.

What is cerebral spinal fluid



An animated picture showing the normal CSF

What are the causes of hydrocephalus?

Hydrocephalus can be congenital, meaning it develops after birth in a short time, mostly due to:

- Inherited genetic conditions that cause the disturbance of CSF flow (like primary aqueduct stenosis)
- Developmental disorders, like brain, spine, or spinal cord birth defects (like Chiari malformation type 2 and or myelomeningocele).
- Premature birth complications(like intraventricular hemorrhage) .
- Maternal infections during pregnancy (like rubella) .



What are the causes of hydrocephalus?

Hydrocephalus can be acquired, meaning it develops at any age, due to a variety of causes, and most commonly due to:

- Central nervous system Infections which are the most common cause of communicating hydrocephalus, like meningitis.
- Trauma or stroke that causes bleeding inside the brain.
- Brain Tumors.



What are the symptoms of hydrocephalus?

In infants, signs and symptoms may include:

Symptoms:

- Vomiting (mostly described as projectile)
- Poor head control
- Developmental delay
- Sucking or feeding problems
- Sleepiness
- Irritability



What are the signs of hydrocephalus?

- An abnormal increase in head size (different than what is known in the family or age group)
- Anterior fontanelle bulge (on the top of the head)
- Fixed downward looking eyes ("sun setting") or are not able to turn outward
- Scalp veins enlargement
- 6th CN palsy
- Cranium enlarges at a rate faster than the facial growth.



What are the signs of hydrocephalus?

Symptoms in older children and adults may include:

- Headache
- Blurred or double vision
- Nausea or vomiting
- Problems with balance
- Slowing or loss of developmental progress like walking or talking
- Decline in school or job performance
- Poor coordination
- Loss of bladder control and/or frequent urination
- Difficulty waking up
- Sleepiness
- Irritability

What are the signs of hydrocephalus?

Symptoms in older children and adults may include:

- Changes in personality or cognition including memory loss
- Walking problems
- Progressive mental impairment
- Dementia
- General slowing
- Poor coordination and balance



How to diagnose hydrocephalus?

Brain scans utilizing both CT and MRI are employed for the diagnosis of congenital and acquired hydrocephalus in individuals of all ages However The initial imaging modality that is used to detect HCP is CT head. These advanced imaging techniques enable medical professionals to:

1. Determine the existence and severity of hydrocephalus by visually inspecting the accumulation of fluid within the brain.

2. Assess for any elevated intracranial pressure (ICP) caused by the buildup of fluid.

 Ascertain whether any underlying structural abnormalities are responsible for or contributing to the development of hydrocephalus.

How to diagnose hydrocephalus?

In certain instances, a routine prenatal ultrasound scan can even detect the presence of congenital hydrocephalus. Early identification permits treatment planning before the baby's delivery.



Head MRI showing dilated ventricles in the left picture (hydrocephalus) compared to normal size ventricles in the right.

Individuals diagnosed with either congenital or acquired hydrocephalus typically require prompt medical intervention to manage the excessive fluid buildup and reduce the intracranial pressure.

Without timely treatment, the increased pressure within the skull can lead to serious and potentially irreversible brain damage. This underscores the importance of early detection and management of hydrocephalus, whether it is present from birth or develops later in life.

There are two primary treatment approaches for both congenital and acquired hydrocephalus:

1. Shunt surgery

Shunt surgery, common surgical management for both acute congenital and acquired hydrocephalus, involves the implantation of a specialized drainage system called a shunt. This thin, tube-like device is placed within the brain to redirect the excess cerebrospinal fluid (CSF) to another area of the body, usually the abdominal cavity, where it can be naturally reabsorbed into the bloodstream. Patients with a shunt can feel the presence of the valve as a small, palpable lump underneath the skin on their scalp. This serves as a visual and tactile indicator of the shunt's placement and functionality



Animated pictures showing the components of VP shunt.

2. Endoscopic third ventriculostomy (ETV)

In addition to shunt surgery, endoscopic third ventriculostomy (ETV) is sometimes employed in the management of hydrocephalus in certain cases.

Patients with obstructive HCP may benefit from ETV. Managing shunt infection may also involve this method (to remove all hardware without putting the patient at risk for elevated ICP). Furthermore, ETV has been suggested as a treatment option for patients who experienced subdural hematomas following shunting (the shunt is removed before to the ETV procedure). Slit ventricle syndrome may also necessitate ETV.



Rather than implanting a shunt system, the ETV procedure involves the surgical creation of a small opening or "stoma" on the floor of the third ventricle, a fluid-filled chamber within the brain. This allows the trapped CSF to bypass the obstruction causing the hydrocephalus and instead drain directly onto the brain's surface, where it can be naturally reabsorbed into the blood.

ETV is particularly well-suited for cases of obstructive hydrocephalus, where the buildup of CSF is primarily due to a blockage in the normal flow and drainage pathways within the brain. By creating this new passage for the fluid, the ETV procedure can effectively alleviate the elevated intracranial pressure without the need for a permanent shunt device

Sources and references:

All pictures used provided by pediatric neurosurgeon consultants at King Fahad University Hospital al Khobar and canva.com choc.org/neuroscience/hydrocephalus/ OpenStax Anatomy and Physiology my.clevelandclinic.org/health/diseases/6008-chiari-malformation nursekey.com/neurologic-and-sensory-disorders/ saintlukeskc.org/health-library/understanding-sixth-nerve-palsy-child kamranaghayev.com/ar/hydrocephalus/ sciencedirect.com/science/article/abs/pii/S1878875017307519 childrens.health.qld.gov.au/health-a-to-z/shunt-surgery nhs.uk/conditions/hydrocephalus/diagnosis/ ninds.nih.gov/health-information/disorders/hydrocephalus

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