Hydrocephalus

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Outline

- 1. Definition, epidemiology, radiographic findings
- 2. Classification of hydrocephalus and etiology
- 3. Clinical presentation
- 4. Physiology of CSF and the blood brain barrier
- 5. Normal Pressure Hydrocephalus

Hydrocephalus

Definition:

An abnormal build-up of cerebrospinal fluid (CSF) in the ventricles of the brain. From the Greek ‘hydro’ (water) and ‘cephalus’ (head)

Incidence:

1 in 500 live births (more common than Down Syndrome!)

Prevalence:

1 to 1.5% of population

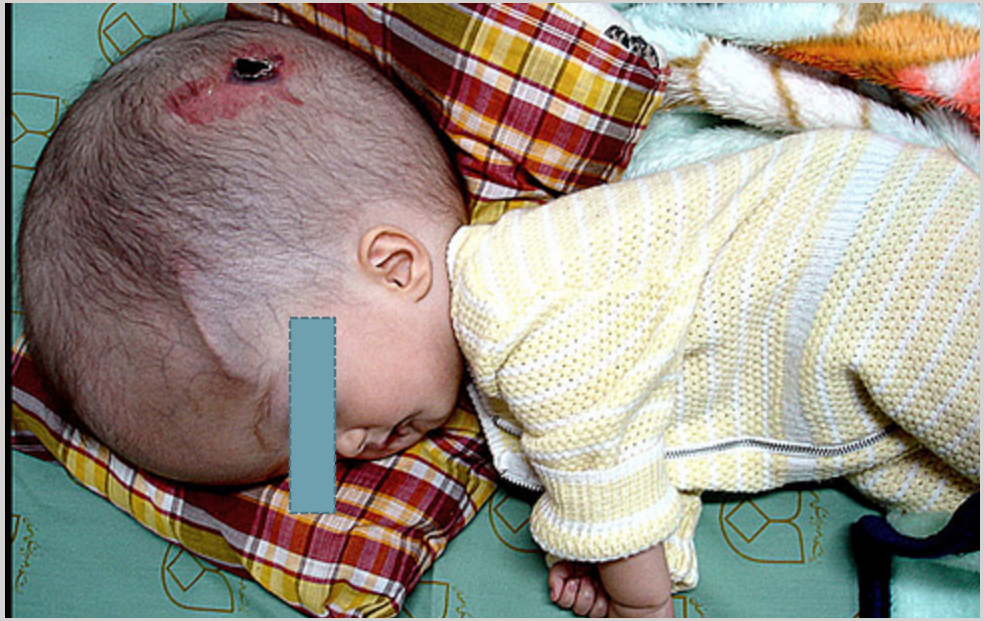
CT/MRI criteria of Hydrocephalus

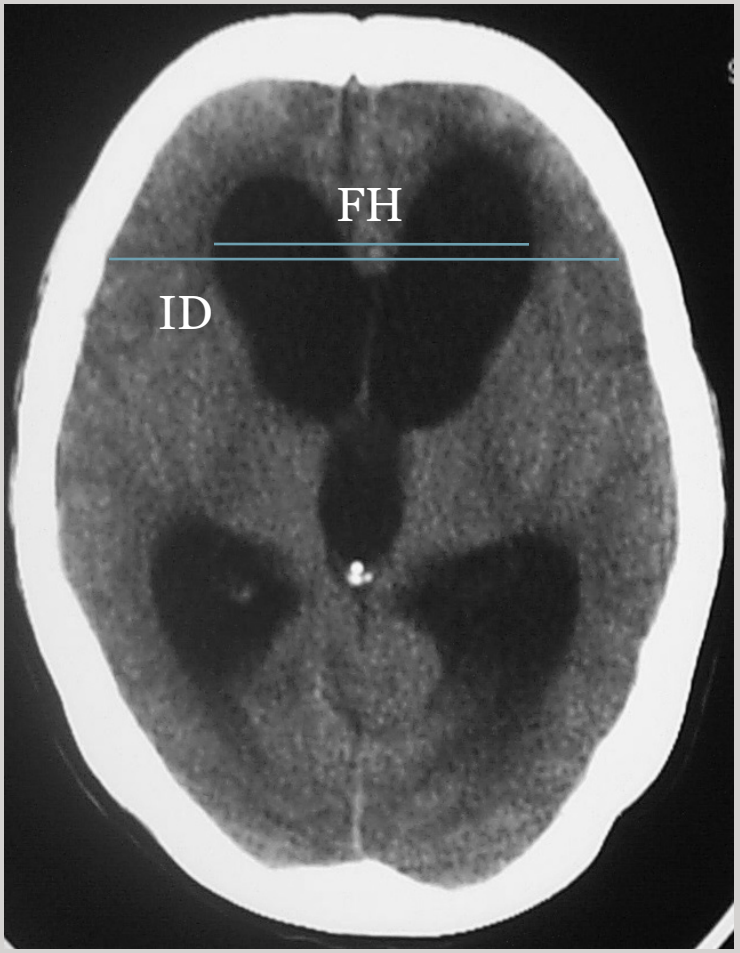
The size of both temporal horns is ≥ 2 mm in width, and the sylvian and interhemispheric fissures and cerebral sulci are not visible

OR

Both TH are ≥ 2 mm and the ratio FH/ID > 0.5 (FH = largest width of frontal horns and ID = the internal diameter from inner-table to inner table at this level)

Hydrocephalus





Other radiographic features of HCP

1. Ballooning of frontal horns and third ventricle
2. Periventricular low density on CT, or periventricular high intensity signal on T2WI on MRI (suggests transependymal absorption)
3. Evan's ratio: ratio of FH to maximal biparietal diameter >30%
4. Sagittal MRI showing upward bowing of the corpus callosum

Functional Classification of Hydrocephalus

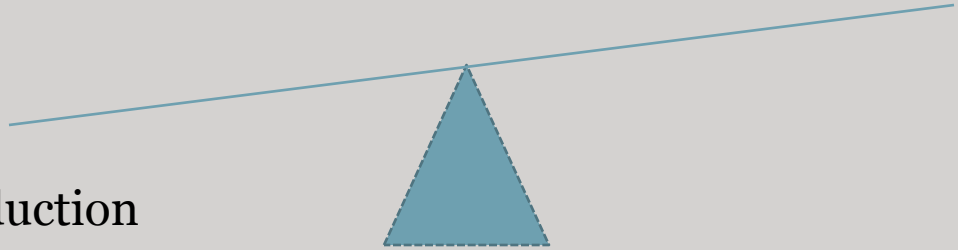
1. **Obstructive** (non-communicating) = block proximal to the arachnoid granulations. Enlargement of ventricles proximal to the block.

2. **Communicating** = CSF circulation blocked at the level of the arachnoid granulations.

Hydrocephalus

Etiology:

Decreased CSF reabsorption



CSF overproduction

Etiology in Pediatric Patients

Congenital (without myelomeningocele) 38%
Congenital (with MM) 29%
Perinatal hemorrhage 11%
Trauma/subarachnoid hemorrhage 4.7%
Tumor 11%
Previous infection 7.6%

Hydrocephalus – Etiology

Congenital:

- Chiari Type 2 malformataion and/or MM
- Chiari Type 1 malformation (4^{th} ventricle outlet obstruction)
- Primary aqueductal stenosis
- Secondary aqueductal gliosis (intrauterine infection or germinal matrix hemorrhage)
- Dandy-Walker malformation (atresia of foramen of Luschka & Magendie)
- Rare X-linked inherited disorder

Hydrocephalus – Etiology

Acquired:

Infectious (the most common cause of communicating HCP)

1. post-meningitis (especially purulent and basal)
2. cysticercosis

Post-hemorrhagic

1. post-SAH
2. post-IVH (20-50% develop permanent HCP)

Secondary to masses

1. non-neoplastic (e.g. Vascular malformation)
2. Neoplastic (medulloblastoma, colloid cyst, tectal glioma, pituitary tumor/apoplexy, choroid plexus papilloma, other tumor around aqueduct)
3. Post posterior fossa tumor removal
4. Neurosarcoidosis
5. Spinal tumor

Post-traumatic

Special forms of Hydrocephalus

1. Hydrocephalus ex vacuo
2. External hydrocephalus
3. Hydranencephaly
4. Normal pressure hydrocephalus
5. Entrapped fourth ventricle
6. Arrested hydrocephalus

Clinical features of active HCP

Young Children:

- cranium enlarges at rate > facial growth
- irritability, poor head control, N/V
- fontanelle full and bulging
- enlargement and engorgement of scalp veins
- Macewen's sign: cracked pot sound on percussing over dilated ventricles
- 6^{th} nerve palsy
- Upward gaze palsy, Parinaud's syndrome
- Hyperactive reflexes
- Irregular respirations with apneic spells
- Splaying of cranial sutures (seen on plain skull X-ray)

Clinical features of active HCP

Older children and adults:

- Headache, N/V
- Gait changes
- Papilledema
- Upgaze and/or abducens palsy

Features of Chronic Hydrocephalus

1. Beaten copper cranium
2. 3rd ventricle herniating into sella
3. Erosion of sella turcica
4. Temporal horns less prominent on CT than in acute HCP
5. Macrocrania: by convention, OFC greater than 98th percentile
6. Atrophy of corpus callosum: best appreciated on sagittal MRI
7. Infants – sutural diastasis, delayed closure of fontanelles, failure to thrive, developmental delay

CSF Physiology

Cerebral cavity capacity of 1600- 1700 mL

150 mL is occupied by CSF

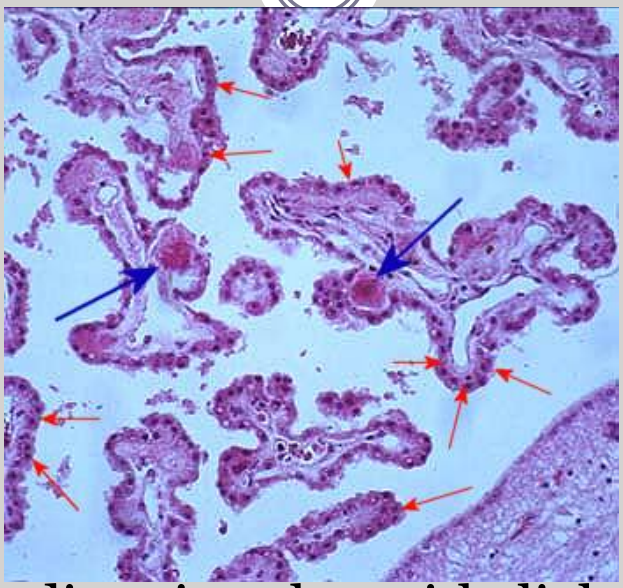
CSF is found in the ventricles, cisterns, and subarachnoid space

Cerebrospinal fluid is formed at a rate of 0.33 cc/min

- 2/3 from choroid plexus
- 1/3 from ependymal surfaces, arachnoidal membranes, perivascular spaces

CSF secretion by the Choroid Plexus

- Choroid plexus is a network of blood vessels lined by a thin layer of epithelial cells.

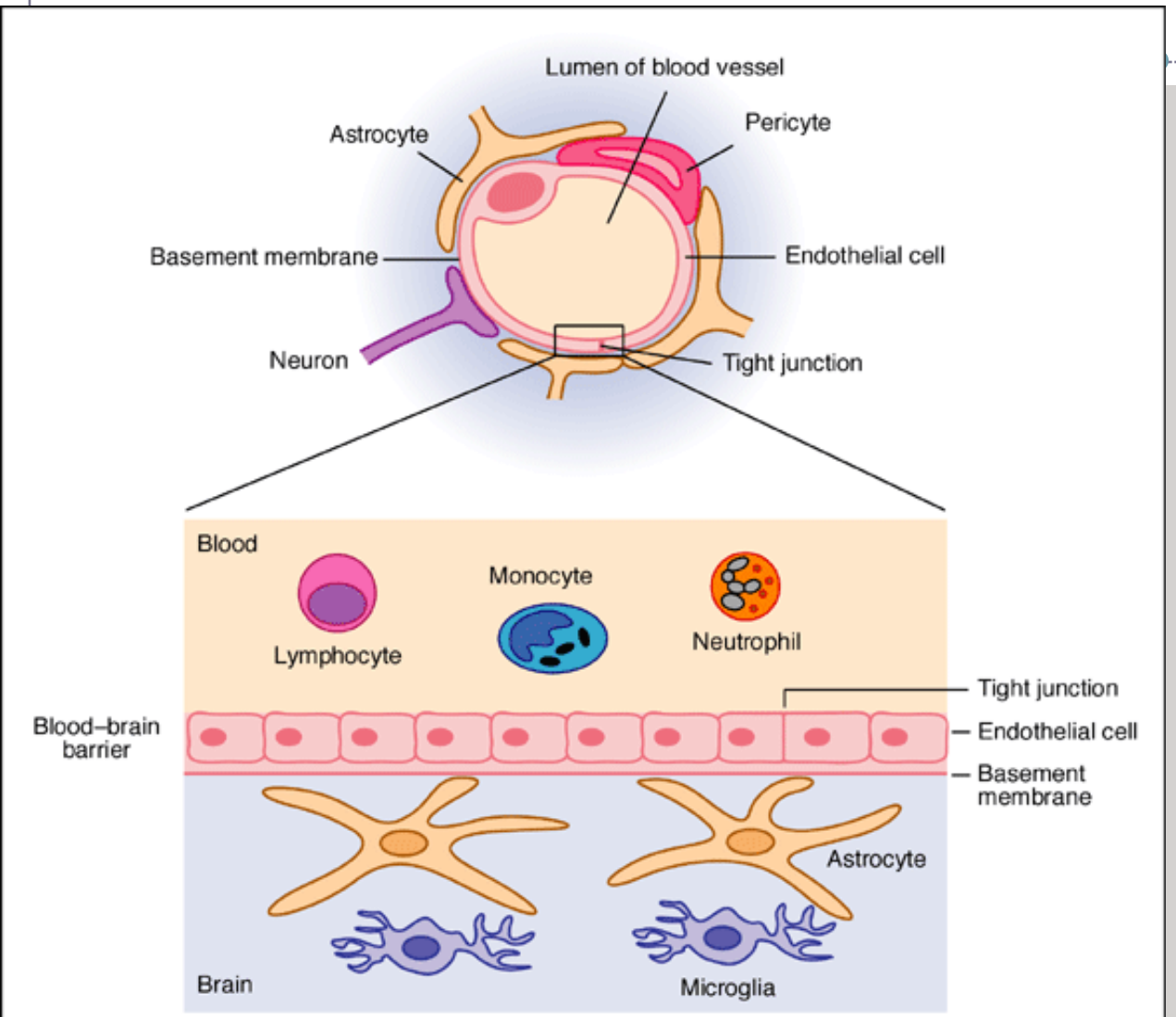


- Active transport of sodium ions by epithelial cells outside of the plexus, draws Chloride. The process depends on the enzyme carbonic anhydrase and can be blocked by the carbonic anhydrase inhibitor Diamox.
- The increase in osmotically active sodium chloride in the CSF causes osmosis of water through the membrane.

CSF flow

Lateral ventricles -> 3rd ventricle -> aqueduct of sylvius -> 4th ventricles -> foramen of Luschka & Magendie -> Cisterna Magna -> subarachnoid space -> arachnoid villi -> venous blood

The Blood Brain Barrier



The blood–brain barrier (BBB)

Expert Reviews in Molecular Medicine © 2003 Cambridge University Press

The barrier exists in the choroid plexus and at the tissue capillaries in all areas except the pineal gland, the subfornical organ the area postrema, the organum vasculosum of the lamina terminalis, some areas of the hypothalamus

Normal Pressure Hydrocephalus

A chronic type of communicating hydrocephalus whereby the increase in (ICP) due to accumulation of cerebrospinal fluid (CSF) becomes stable and that the formation of CSF equilibrates with absorption. The ICP gradually falls but still maintains a slightly elevated level and the CSF pressure reaches a high normal level of 150 to 200 mmH_2O.

Normal Pressure Hydrocephalus

Clinical Features:

Age of presentation usually >60 y/o, slight male predominance

Gait disturbance – wide based, short shuffling steps, unsteadiness on turning, feel like glued to the floor, difficulty initiating steps or turn

Dementia – apathy, dullness in thinking, inattention (thought to be due to traction on limbic or

Incontinence (a late finding) – increased urinary frequency + urgency

Normal pressure on random LP

Symptoms remediable with CSF shunting

Normal Pressure Hydrocephalus

CT/MRI features:

1. communicating hydrocephalus
2. transependymal CSF absorption
3. compression of convexity sulci
4. rounding of the frontal horns

Normal Presssure Hydrocephalus

Lumbar puncture:

OP> 100 mmH2O have a higher response rate to shunting.

Ambulatory lumbar drainage:

High positive predictive value of response to drainage of 300 cc/day and clinical improvement with 5 day trial.

Radionuclide cisternography:

Persistence of ventricular radionulcied activity 48-72 hrs after administration of marker correlates with 75% chance of improvement with shunting

Patients who clear over 50% of total intracranial radioactivity within 24hrs are unlikely to improve with shunting.

Normal Pressure Hydrocephalus

Treatment:

Dementia work-up prior to surgical procedure

VP shunt with medium pressure valve (closing pressure 65-90 mmH2O)

Gradually sit patient up over a period of several days; proceed more slowly if develop low pressure headache

Follow with CT for 6-12 months

Evaluate for shunt malfunction if no improvement or change in ventricle size. If not obstructed may try a lower pressure valve.

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