Hydrocephalus

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Hydrocephalus

- Derived from Greek words:
  - "Hydro" – water
  - "Cephalus" – head
- As the name implies, it refers to a general condition whereby there is excess Cerebro-Spinal Fluid (CSF) within the intracranial space and, specifically, the intraventricular spaces within the brain…causing dilation of the ventricles, and a wide range of symptoms.

Facts and Physiology

- The majority of CSF is produced by the **choroid plexus** within the brain through a metabolically active process (i.e. requires ATP) whereby sodium is pumped into the subarachnoid space, and water follows from the blood vessels.
- Choroid plexus is primarily located in the lateral ventricles (temporal horn roofs, and floors of bodies), posterior 3rd ventricle roof, and caudal 4th ventricle roof.

Facts and Physiology

- The average adult brain produces between 450 and 600 cc's of CSF every day.
  - Normally, production = resorption.
- At any given moment, there is only ~150 cc's of CSF present in the average adult; of this, only ~25 cc's is within the brain ventricles!
  - So, the CSF volume turns over three to four times every day, with only a very small fraction of the CSF being in the ventricles at any given time, even though the majority of it is produced there!

Important concept to build on

- The CSF production/resorption system exists in a delicate balance under normal conditions.

The CSF pathway

- From the lateral ventricle (remember, there are two…one on each side), CSF travels through the foramen of Monro into the 3rd ventricle (midline), then passes through the Cerebral Aqueduct [of Sylvius] into the 4th ventricle. It then exits the 4th ventricle through either of two Foramina of Luschka ("L" is for Lateral/Luschka) or the single Foramen of Magendie ("M" is for Midline/Magendie).

CSF pathway (cont'd)

- After exiting the 4th ventricle, the CSF flows through the subarachnoid space over and around the brain and spinal cord, and is eventually reabsorbed into the venous (blood) system through numerous arachnoid granulations along the dural venous sinuses (especially the superior sagittal sinus).

CSF pathway (cont'd)

- Arachnoid granulations contain arachnoid villi, which function as pressure-dependent one-way valves that open when the ICP is ~3 to 5 cm H20 greater than dural venous sinus pressure.
- Remember – CSF production is a metabolically active process which uses ATP, whereas CSF resorption is a passive process that is driven by the pressure gradient between the intracranial space (ICP) and the venous system (~CVP).

Hydrocephalus

- Two major distinctions:
  - Communicating Hydrocephalus (CoH)
    - Also known as “non-obstructive” hydrocephalus
  - Non-communicating Hydrocephalus (NCH)
    - Also known as “obstructive” hydrocephalus

Hydrocephalus terminology

- It is important to understand the difference between "Communicating" and "Non-communicating" hydrocephalus.
  - If the CSF pathway is "open from start to finish," meaning CSF can travel freely from the choroid plexus to the arachnoid granulations, then you have "no obstruction" and a *communicating* hydrocephalus.
  - If the CSF can't travel freely from start to finish, then you've got an "obstruction" and *non-communicating* hydrocephalus.

Let's look first at Communicating Hydrocephalus…

CoH

- In the vast, vast majority of cases, this represents a problem with CSF resorption; simply put, it cannot keep the pace with CSF production.
  - Normal CSF production = resorption
  - Comm. Hydrocephalus CSF production > resorption
- As a result, the ventricular system dilates uniformly, and ICP rises.

Still building…

- Remember that the normal system exists in a fairly delicate balance. A major insult is not required to upset this balance; sometimes, a very slight disturbance is enough to tip the scales such that, over time, communicating hydrocephalus results.

CoH

- Very rarely, there is overproduction of CSF (rather than under-absorption) which leads to disruption of this balance, and development of communicating hydrocephalus.
- This is rare, but Choroid Plexus Papillomas have been known to present this way.

CoH

- Signs/Symptoms
  - In young children whose cranial sutures have not yet fused, you can see disproportional increase in head circumference compared to the rest of the face/body.
  - In children with fused sutures/adults, hydrocephalus manifests with symptoms of increased intracranial pressure;
    - H/A, N/V, papilledema, gait disturbance, 6th cranial nerve palsy, upgaze difficulty, etc.

CoH etiology

- Infection (incidence after bacterial meningitis can approach 30%)
- Subarachnoid Hemorrhage (blood and blood breakdown products cause scarring of arachnoid granulations)
- Post-operative
- Head trauma
- Etc.

Still building…

- Although we usually think of Communicating Hydrocephalus as being a "gentle" disruption of the balance between CSF production and resorption that occurs over time, sometimes there can be a sizeable insult that causes an acute disruption of the balance, and CSF resorption is suddenly (and dramatically) reduced.

Still building…

- Given the high turnover of CSF in the normal individual, you should quickly recognize that a dramatic decline in CSF resorption ability can represent an emergency. If a significant number of arachnoid granulations are impaired, say, by subarachnoid blood (such as from a ruptured aneurysm), hydrocephalus can develop very quickly. Neurological decline in such a situation can be rapid, with patients becoming sleepy, then obtunded, then requiring intubation.

Let's pause the CoH discussion, now, and turn our focus to Non-communicating Hydrocephalus (NCH)

NCH

- Non-communicating hydrocephalus occurs whenever there is ANY physical obstruction to the normal flow of CSF before it leaves the ventricles.

NCH causes

- Many, many causes:
  - Aqueductal stenosis
  - Tumors/Cancers/Masses
  - Cysts
  - Infection
  - Hemorrhage/hematoma
  - Congenital malformations/conditions
  - Etc.

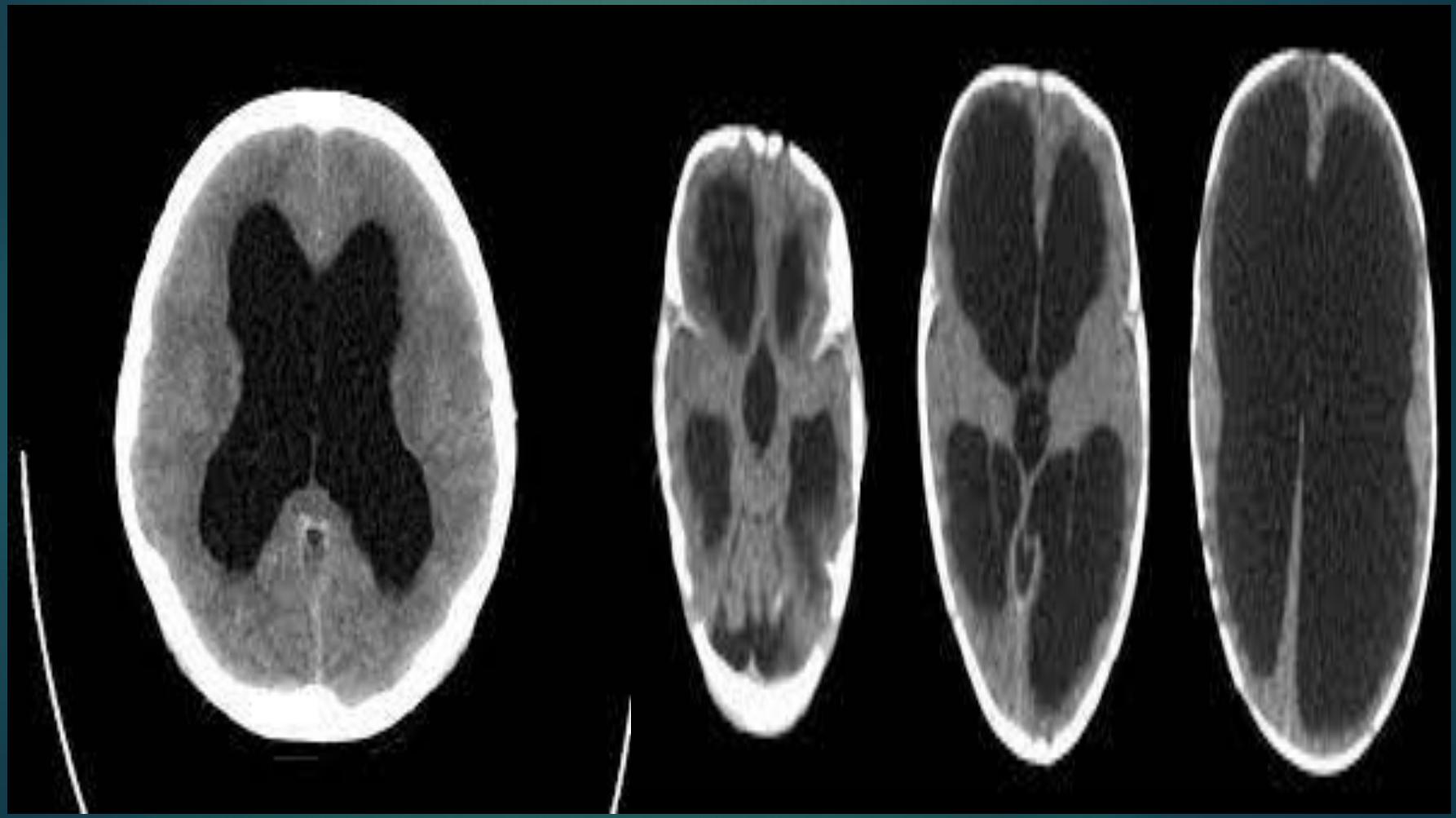
The bottom line…NCH

- Non-communicating hydrocephalus can result from CSF flow obstruction at any point along the intraventricular pathway.
- Processes that have developed over longer periods of time (i.e. masses, etc) usually cause gradual development of symptoms.
- Acute processes (i.e. intraventricular bleed) can cause acute obstruction with rapid mental status decline (just like we discussed previously).

Radiography

- The earliest consistent radiographical finding indicative of development of hydrocephalus is dilation of the temporal horns of the lateral ventricles (arrows). In most younger and middle-aged patients, these should be almost invisible.

This is an abnormal scan!



Hydrocephalus treatment

- Treatment for Hydrocephalus remains surgical.
  - Acute hydrocephalus, whether communicating or not, usually necessitates urgent or emergent placement of an External Ventricular Drain (EVD - catheter passed through the patient's scalp and skull into lateral ventricle, that drains CSF to a collection system kept at the patient's bedside).
    - EVD cannot be maintained indefinitely; usually, if the patient is unable to tolerate weaning/clamping of the EVD prior to removal, a permanent shunt will be required.

Hydrocephalus treatment

- For communicating hydrocephalus, the mainstay of treatment is shunt placement.
  - Ventriculo-peritoneal is most used.
  - Lumbar-peritoneal sometimes utilized, though overdrainage is a problem.
  - Acute communicating hydrocephalus patients (i.e. SAH) can sometimes be managed with EVD with successful weaning and no shunt placement, but a significant number of these patients eventually need shunt placement weeks or months later.

Non-communicating Hydrocephalus

- NCH treatment also surgical, but sometimes shunt can be avoided by removing the obstructing lesion.
  - Colloid cyst at anterior 3rd ventricle causing ball/valve obstruction of Foramen of Monro.
  - Pineal region tumor causing compression of cerebral aqueduct.
  - Ependymoma blocking 4th ventricular CSF outlets.
  - Many, many more!

NCH treatment

- In addition to surgical removal of obstructive lesion and shunt placement, Third Ventriculostomy is an option for non-communicating hydrocephalus.
  - Often performed in conjunction with VP shunt placement.
  - Hole is surgically opened in floor of third ventricle so CSF flows out into the interpeduncular cistern and pre-pontine space (bypasses cerebral aqueduct).

Previously shunted patients

- Take note: Every patient with a history of VP shunt placement who presents with a headache is NOT automatically having a shunt malfunction/infection.
  - Yes, you should consider that in your differential, but still work it up appropriately. After all, the vast majority of patients with headaches don't have VP shunts.

Previously shunted patients

- Nevertheless, VP shunts can and do fail for any of a number of reasons.
  - The incidence of shunt failure is as much as 40% the first year (most in the first few months), and then 5% per year after the first year.
  - 50% of shunts fail by 5 years from placement.
    - Mechanical failure from occlusion/disconnection, migration, overdrainage/underdrainage, infection, skin erosion, etc.

One last thing to discuss…

Normal Pressure Hydrocephalus

- Normal Pressure Hydrocephalus (NPH) - This is a distinct clinical entity that was first described in 1965.
- This is important and you MUST know it because it is one of the rare preventable and/or reversible causes of dementia.
  - Under-recognition leads many patients with NPH to be diagnosed with Alzheimer's or age-related dementia and they never seek (or receive) treatment.

NPH

- **Classic triad: "Wet, Wobbly, and Wacky!"**
  - Urinary incontinence
  - Gait disturbance (usually the first symptom to present) – wide stance; short, shuffling steps
  - Rather quickly-progressive dementia
- Communicating hydrocephalus on CT/MRI
- Lumbar Puncture (LP)
  - Normal opening pressure
  - Symptoms improve with CSF removal

NPH treatment

- Procedure of choice for patients felt to be suffering from NPH is VP shunt placement.
  - LP shunts tend to overdrain.

NPH prognosis

- Chance of outcome is improved if symptoms have been present for shorter period of time
  - Meaning that failure to recognize these patients delays their treatment and lessens their chances!
- Least likely symptom to improve with shunting is dementia
  - It's always a worse sign once mentation gets involved!
- Most likely symptom to improve is incontinence

NPH: Bottom Line

- Good history taking and time with patients and family is key to nailing this diagnosis. Just because they've got the classic triad doesn't mean it may not be Alzheimer's (it often will be); but if you keep NPH on your differential and consider it rather than writing off all dementia as Alzheimer's, you will have the opportunity to make a fantastic difference in the lives of many patients.