Update on Hydrocephalus Management

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Disclosures

- Nothing to disclose

Learning Objectives

- Describe the pathophysiology of hydrocephalus.
- List common etiologies of hydrocephalus in the pediatric patient.
- Discuss key treatment strategies in the management of hydrocephalus.
- Describe shunt complications in the pediatric patient.
- Describe ideal primary care surveillance/management of a shunted patient.

Plan for Today

- Definitions
- Anatomy/pathophysiology
- Etiologies
- Clinical Presentation
- Treatment Options
  - Endoscopy and Shunts
  - Surveillance

Hydrocephalus

Abnormal accumulation of cerebrospinal fluid (CSF) in the brain

- CSF continuously made in the choroid plexus of the ventricles
- Production is independent of pressure
- Regulated by enzymes – certain meds can turn down production, but not off

CSF and the Ventricles

Circulates through the ventricular system

- right and left lateral ventricles
- Foramen of Monro
- 3rd ventricle
- Cerebral aqueduct
- 4th ventricle

Leaves ventricles by foramina of Luschka & Magendie and percolates around brain and spinal cord (subarachnoid spaces)
CSF Pathways

- CSF circulates over surface of brain where it is reabsorbed back into the veins of the bloodstream
- Arachnoid granulations

Pathophysiology

- CSF production ➔ CSF absorption
- Mechanisms leading to hydrocephalus
  - Block flow of CSF
  - Block absorption of CSF
  - Overproduction of CSF (rare)
- “Blockage” varies by location and nature
- **Obstructive Hydrocephalus**: Intraventricular Block
- **Communicating Hydrocephalus**: Absorption Failure

Types of Hydrocephalus

- **Congenital**
  - most structural/genetic
  - some prenatal events (ie. acquired, but prenatal)
  - Ex: Spina Bifida, Dandy-Walker, CMV
- **Acquired**
  - post-hemorrhagic
  - post-inflammatory
  - post-infectious
  - Ex: IVH, Tumor, Neonatal Meningitis

Obstruction to CSF flow at Cerebral Aqueduct

- Aqueductal stenosis
- Atresia
- Webs, bands
- Scarring (gliosis) after infection or hemorrhage
- Mass lesions (tumors, cysts)

Aqueductal Stenosis

- Normal Aqueduct
- Web
- Stenosis
- Atresia
X-linked hydrocephalus

- Outlets of 4th ventricle blocked
- Dandy-Walker cyst
- Infection, hemorrhage
- Mass lesion
- Chiari I malformation

Obstruction of CSF flow at level of 4th ventricle

- Cystic enlargement of 4th ventricle
- 70-90% of patients develop hydrocephalus
- Cyst blocks cerebral aqueduct and/or 4th ventricular outlets → hydrocephalus
- Sometimes need to shunt ventricles and/or cyst
- Associated with other central nervous system and systemic abnormalities

Dandy-Walker Cysts

Dandy-Walker Cysts

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Example of Obstructive Hydrocephalus

- “Communicating hydrocephalus”
- Hydrocephalus and myelomeningocele/Chiari II malformations
- Scarring of arachnoid granulations
- Infection, inflammation, hemorrhage
- Post-hemorrhagic hydrocephalus of prematurity

Blockage of CSF flow in subarachnoid space

Posterior fossa tumor causing obstructive hydrocephalus
Spina bifida/Myelomeningocele

- 80-90% of MM patients have treatable hydrocephalus
- Caused by obstruction by Chiari and underdevelopment of cortical subarachnoid spaces

Hydrocephalus and the Premature Newborn

- Germinal matrix hemorrhage ≤ 32 weeks
- Maximum risk ~ 23 weeks gestation
- Bleed can extend into ventricles, parenchyma
- Grade I-IV depending on severity, extent of hemorrhage, presence of dilated ventricles
- Blood → obstructs CSF pathways
- Blood breakdown products → scarring along subarachnoid space, arachnoid villi → impedes CSF absorption → hydrocephalus

Germinal Matrix Hemorrhages in Premature Infants

Grade I    Grade II    Grade III    Grade IV

Example of IVH

Other Reasons for Shunts

- Arachnoid Cyst
- CSF leak
- Pseudotumor Cerebri
- Non-Accidental trauma

Clinical Presentation in Infants

- No signs or symptoms
- Irritability
- Vomiting
- Large head/accelerated head growth – crossing percentiles
- Full fontanelle
- Poor head control
- Increased LE spasticity
- Enlargement/engorgement of scalp veins
- Eye crossing, jiggling eyes
  - Cranial nerve VI palsy
  - Sun setting sign
- Split sutures
- Bradycardia, apnea
- Delayed milestones
Clinical Presentation in Older Children
- Headache
- Nausea, vomiting
- Irritability, lethargy
- Papilledema
- Vision changes – crossing eyes, upgaze palsy, double or blurred vision, blindness
- Neck pain
- Gait changes
- School / learning / cognitive problems

Upgaze & Bilateral CN VI Palsies

Fundoscopic Exam
- Normal exam
- Papilledema

Treatment Options
- Drugs
- Serial Lumbar Puncture
- Serial Ventricular Puncture
- External ventricular drainage
- Reservoir/ventricular access device (VAD)
- Ventriculostomy
- Ventriculoperitoneal (VA, VPI) shunt
- Endoscopic Third Ventriculostomy (ETV)

Shunt Basics
- 3 Parts: Proximal Catheter(s), Valve & Distal Tubing
  - VP, VPI, VA, Cystoperitoneal, Others

Management of the Preemie with Hydrocephalus after IVH
- Serial lumbar punctures; eventual dry taps
- Ventricular taps; develop porencephaly
- Surgical placement of ventricular reservoir for daily ventricular aspirations
- Ventriculo-subgaleal shunt
Shunt Problems

- Symptoms of increased ICP
  - HA, irritability, lethargy, vomiting, bulging fontanel, increasing OFC velocity, seizures, down-gazing eyes, decline in school performance
- Shunt Tap
- Imaging
  - CT vs MRI
  - About 25% of pts do not enlarge their ventricles with malfunction!!

Shunt Malfunction & Post Shunt Revision

- Shunt malfunction
  - Usually due to proximal catheter obstruction
    - Rare for valve or peritoneal catheter to be clogged
    - Clogged peritoneal catheter suggests infection
  - Ventricles may or may not enlarge!
  - Can see ventricular enlargement with constipation, seizures, UTI

Shunt Infection

- ~5% risk of early infection after shunt surgery
- Usually occur within 6 months of shunt surgery
- *Staph epidermidis* from skin most common pathogen
- Blood work – CRP, ESR usually elevated
- Shunt tap
- Shunt needs to be removed, external ventricular drain placed, long-term abx, replace shunt once CSF sterile
  - Duration “on drainage” depends on organism and ongoing lab work

External Ventricular Drain

- Abdominal pseudocyst
  - Usually represents an indolent infection
  - Can present as
    - Shunt malfunction
    - Abdominal mass, pain
  - History of recent abdominal surgery
  - Drain pseudocyst, move shunt if not infected
  - If infected, externalize shunt, replace after CSF clear
Shunt Disconnection

Endoscopic Third Ventriculostomy

- ETV creates a hole connecting the third ventricle to the subarachnoid space
- Helpful for some patients with aqueductal stenosis, 20% may still require shunting
  - Not for pts with communicating hydrocephalus
  - Likelihood of success depends on several factors
    - Age, shunt history, etiology of hydrocephalus

Managing a child with Hydrocephalus

- Neurosurgery follow-up important, identify a contact at the patient’s neurosurgery department.
- Why was the shunt placed?
- What kind of valve (programmable? setting?)
- What does child look like during a shunt malfunction?
- Copies of scans on file for local ER/ future comparison
- Do ventricles get large when shunt malfunctions?

Programmable valves?

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Summary

- Hydrocephalus can occur in isolation or in association with other conditions
- Different causes, mechanisms, and etiologies
  - Obstruction to flow in the ventricles ("Non-communicating")
  - Obstruction at level of subarachnoid spaces / arachnoid villi ("Communicating")
- Presentation varies by age
- Good treatments, not perfect
  - Be pro-active and form relationships with colleagues
References