

# CHOC Macrocephaly/Hydrocephalus August 25, 2021, 12:30 – 1:30 PM (PST)

August 25, 2021, 12:30 - 1:30 PM (PST)





#### **Hydrocephalus**

Hydro - water.

Kephale - head.

Condition secondarily to obstruction of the cerebrospinal fluid pathways, and accompanied by an accumulation of cerebrospinal fluid within the skull; The fluid is usually under increased pressure, but occasionally may be normal or nearly so. It is typically characterized by enlargement of the head, prominence of the forehead, mental deterioration, and convulsions; May be congenital or acquired.



#### **Hydrocephalus**

- Pathological hydrocephalus: usually an emergency, patient needs to see a neurosurgeon
- Benign hydrocephalus: patient does not need a neurosurgeon, parents of child invariably want to see a neurosurgeon



#### **Anatomy of CSF Pathways**





# Advances in the treatment of hydrocephalus

1950-1995: Standard of care usually involved implantation of ventriculo-peritoneal shunt.

1995-present: Standard of care has migrated to endoscopic third ventriculostomy, without implanted hardware if possible.

1999-present: Computer and magnetically controlled shunts implanted in shunt-dependent patients.



#### **Signs and Symptoms**

Large (growing) head Circumference **Bulging, firm anterior fontanelle Dilated scalp veins Ocular palsies ("Sun downing") Irritability** Lethargy Failure to thrive, vomiting, weight loss Bradycardia, hypertension, respiratory irregularities





#### Hydrocephalus (from Hess, 1922)







#### **CT Scan: Ventriculomegaly**







#### **Grade IV PIVH**



#### **Post-hemorrhagic Hydrocephalus**







#### Grade IV PIVH: pre- & post-op



CHOC-

#### **Complex PHH-trapped V<sub>4</sub>**









## AKA (often erroneously)

- Benign extracerebral fluid collections
- Benign enlargement of subarachnoid fluid spaces
- External hydrocephalus
- Benign familial macrocephaly
- Benign macrocephaly
- Subdural hygromas
- Pseudohydrocephalus



### **Clinical Findings**

- Head circumference >95%ile
- Head growth has crossed >2 isobar percentile lines
- Flat or sunken anterior fontanelle
- Frontal bossing
- Family history of macrocephaly





Infants with Birth Occipitofrontal Circumferences (OFC) Greater than 95th Percentile 8 Boys, 8 Girls



## **Primary Etiology**

- Cerebral/meningeal congestion
- Defect of CSF absorption
- Increased resistance at arachnoid villi of the superior sagittal sinus
- May be autosomal dominant trait

#### **CSF** Absorption





## **Secondary Etiology**

- Primary neonatal subarachnoid hemorrhage
- Birth trauma
- Congenital heart disease

#### **Developmental History**

- Transient delay at 5-12 months
- Normal by 15-24 months of age
- Long-term head circumference stabilizes at 90<sup>th</sup>%ile





BOYS

#### **Associated Conditions**

- Posterior plagiocephaly/benign positional molding
- Sawin, Muhonen, et al, Journal of Neurosurgery, May 1996



#### Brachycephaly: Bioccipital Plagiocephaly









#### **Associated Conditions**

Extra-axial bleeding "Pseudo" subdural bleeding Retinal hemorrhage/Tersons syndrome



#### 12 17 AM A 9

0 5

/ 90

SP FoV 172

Corst





PM

#### Treatment

- Expectant observation
- Elevate head of bed
- Rarely: Shunting device (done by neurosurgeons not familiar with long-term conservative treatment)
- Controversial: Acetazolamide/furosamide

## Medications that decrease CSF production:

Acetazolamide (Diamox)

**Furosamide (Lasix)** 

Used together, both may decrease CSF production by 50% Side effect: metabolic acidosis


#### **Benign Communicating Hydrocephalus of Infancy**

#### **Radiographic History**

- Subarachnoid collections persist until 18-24 months of age
- Normal CT at 2.5 years of age







#### Clues to differentiate shaken baby syndrome from BCHI

- Associated injury: Bruises, old fractures
- Skull fractures
- Brain parenchyma injury
- Social situation risks

#### **Big head = less movement**

#### Familial Macrocephaly







Shunts are associated with a higher complication rate than any other procedure in Neurosurgery

- Infections
- Occlusion
- Over and under drainage
- Replaced on average 10x in lifetime



#### **Proximal Malfunction**





#### **Distal Malfunction**



CHOC-





#### **Lost Catheter - Brain**



CHOC-

#### Migration into Subgaleal Space







#### Migration into Subgaleal Space







#### Migration into Subgaleal Space





#### **Overdrainage (ghost brain...)**





#### **Ove<u>rdrainage - Subd</u>ural**











#### **Migration into Head**







# Many shunt problems arise out of poor (surgical) planning...



#### **Sea Serpent**





#### Beaded and Flanged Catheters











#### Zur falschen Zeit am falschen Ort im falschen Job?

### Shunts do not fit well inside tiny babies...

#### **Decubitus**





#### **Calcification**









#### **Fracture Along Tract**







## **Correction of Spinal Deformity**





#### Migration into Chest Pleural effusion





#### **Pseudocysts**



CHOC-





#### Hernia





#### **Distal Knot**





#### **Distal Knot**





#### **Too Many Shunts**











#### **Extra-Peritoneal**








## **Third Ventriculosotmy:** Alternative Treatment for Obstructive Hydrocephalus

- Redirection of CSF Pathway
- Non-communicating hydrocephalus
- Preservation of subarachnoid to venous pathway
- "Non-infantile/acquired obstruction"





## **Third Ventriculostomy:** Alternative treatment for obstructive hydrocephalus



Fig. 15. Endoscopic view into right anterior horn. (lenscope)
1: choroid plexus
2: foramen of Monro
3: septal vein
4: thalamostriate vein





Fig. 17a. Endoscopic view through the foramen of Monro into the anterior part of the third ventricle. (fiberscope) IR: infundibular recess OC: optic chiasm LT: lamina terminalis CA: anterior commissure large arrow: mamillary bodies asterisk: tuber cinereum

Fig. 17b. Endoscopic view through the foramen of Monro into the anterior part of the third ventricle. (lenscope) C: optic chiasm small asterisk: mamillary bodies large, open asterisk: tuber cinereum (note the bluish colour) arrow: infundibular recess













## In closing, what is it?



CHOC-



## THANK YOU



