Hydrocephalus in Children: Diagnostic Imaging and Radiological Characteristics

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Agenda

- Introduction to the Patient
- Macrocephaly
- Basics of Hydrocephalus
- Diagnostic Imaging of Hydrocephalus
  - Ultrasound, CT, MR
- Shunts
The Patient

- 6 yo boy with macrocephaly, which manifested in the first year of life as excessive head growth, accompanied by reduced activity and poor feeding.
- Other significant medical hx includes cognitive delay, osteogenesis imperfecta type 2, restrictive lung disease.
- Birth hx: born at 34 ½ wk premature by c-section with subsequent 25 day NICU course significant for apneic and bradycardic spells
- No family hx of large heads

Used with permission of patient’s mother
Macrocephaly in Infant or Child

- Defined as a head circumference more than 2 SD above the mean for age and sex
- Excessive rate of head growth over time suggests increased intracranial pressure
  - Most often caused by hydrocephalus, extra-axial fluid collections, or neoplasms
- Macrocephaly with normal head growth rate suggests familial macrocephaly or true megalencephaly
## DDx of Macrocephaly

<table>
<thead>
<tr>
<th>Causes</th>
<th>Examples</th>
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<tbody>
<tr>
<td>Pseudomacrocephaly, pseudohydrocephalus, catch-up growth</td>
<td>Growing premature infant, recovery from malnutrition, congenital heart disease</td>
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<tr>
<td>Increased intracranial pressure with dilated ventricles with other mass</td>
<td>Progressive hydrocephalus, subdural effusion, hydrancephaly Arachnoid cyst, porencephalic cyst, brain tumor</td>
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<tr>
<td>Benign familial macrocephaly (idiopathic external hydrocephalus)</td>
<td>Benign enlargement of subarachnoid spaces, congenital communicating hydrocephalus, benign subdural collections of infancy</td>
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<tr>
<td>Megalencephaly</td>
<td>Benign familial (see above)</td>
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<tr>
<td>with neurocutaneous disorder</td>
<td>Neurofibromatosis, tuberous sclerosis</td>
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<td>with gigantism</td>
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<td>with dwarfism</td>
<td>Achondroplasia</td>
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<tr>
<td>metabolic</td>
<td>Mucopolysaccharidoses, Krabbe’s disease, Ganglioside storage disease</td>
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<td>lysosomal</td>
<td>Metachromatic leukodystrophy</td>
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<td>other leukodystrophy</td>
<td>Canavan spongy degeneration</td>
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<td>Thickened skull</td>
<td>Fibrous dysplasia (bone), hemolytic anemia (marrow), sicklemia, thalasemia</td>
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Workup of Macrocephaly

- **History**
  - including family medical hx

- **Physical exam**
  - Neurological exam
  - Plotting measurement of head circumference for age and sex
  - Palpation of anterior fontanelle (if not closed) and of head for asymmetries or ridges
  - Listen for bruits in neck and head
  - In infants, transillumination of skull in a darkened room may reveal subdural effusions, hydrocephalus, hydranencephaly, or cystic defect

- **Imaging studies**

- **Depending on hx, possible labs include** toxoplasmosis serum antibody levels, lumbar puncture, subdural tap
Narrowed DDx for our Patient

- Hydrocephalus
- Space-occupying lesion (e.g., tumor or cyst)
- Extra-axial fluid collection (e.g., subdural effusion)
- Growing premature infant
Pathophysiology of Hydrocephalus

- Imbalance of CSF formation and absorption, resulting in an excess of CSF with subsequent increase in intracranial pressure

- CSF basics
  - Normal CSF production 0.2-0.35 mL/min with a majority produced by the choroid plexus
  - Total volume of CSF in an adult ~ 120 mL
CSF Circulation & Ventricular System

Epidemiology

- Incidence of congenital hydrocephalus is 2-3 per 1,000 live births
- Incidence of acquired hydrocephalus is not known
- About 100,000 shunts are implanted each year in the developed countries
- Incidence is equal in males and females
Symptoms of Hydrocephalus in Infants

- Poor feeding
- Irritability
- Reduced activity
- Vomiting
Signs in Infants

- Macrocephaly with excessive rate of head growth
- Dysjunction of sutures
- Dilated scalp veins
- Tense/bulging fontanelle
- Setting-sun sign: Eyes are deviated downward, the upper lids are retracted, and superior sclerae may be visible.
- Lower limb spasticity and hypertonia
- Papilledema often not present in infants
Symptoms in Children

- Altered behavior
- Slowing of mental capacity or decreased level of consciousness
- Headaches (initially in AM)
- Neck pain (2º to tonsilalar herniation)
- Vomiting (worse in AM)
- Blurred vision (2º to papilledema)
- Double vision (2º to Abducens nerve palsy)
- Stunted growth and sexual maturation from third ventricle dilatation → obesity, precocious or delayed onset of puberty
- Difficulty in walking 2º to spasticity
Signs in Children

- Papilledema → optic atrophy and vision loss (if increased intracranial pressure (ICP) goes untreated)
- Failure of upward gaze (2º to pressure on tectal plate through the suprapineal recess)
- Macewen sign: A "cracked pot" sound is noted on percussion of the head.
- Unsteady gait (2º to spasticity in lower extremities)
- Macrocephaly (sutures are closed, but chronic ICP will lead to progressive abnormal head growth)
- Uni- or bilateral sixth nerve palsy
Classifications of Hydrocephalus

- Congenital v. Acquired
- Communicating v. Noncommunicating
Congenital Hydrocephalus

- Most common category in children
- Usually present during infancy
  - Hydrocephalus presenting after age 6 months is less likely to be congenital, and neoplasm must be excluded
Congenital Causes

- **Aqueduct (of Sylvius) stenosis**
  - due to malformation (10% of all cases in newborns)
    - Arnold-Chiari I & II, Vein of Galen, Klippel-Feil
    - Postinfectious: toxoplasmosis, cytomegalic inclusion disease, rubella, syphilis

- **Obstruction of foramina of Luschka and Magendie**
  - Dandy-Walker malformation (2-4% of newborns)

- **Agenesis of foramen of Monro**

- **Bickers-Adams syndrome**

- **Achondroplasia**
Acquired Causes

- Mass lesions
  - account for 20% of all cases in children
  - usually tumors (eg, medulloblastoma, astrocytoma), but cysts, abscesses, or hematoma also cause
- Intraventricular hemorrhage
  - 2º to prematurity, head injury, or rupture of a vascular malformation.
- Infections
  - Meningitis, especially bacterial, Mumps, cysticercosis
- Increased venous sinus pressure
  - 2º to achondroplasia, craniostenoses, or venous thrombosis.
- Iatrogenic
  - Hypervitaminosis A.
- Idiopathic
Communicating Hydrocephalus

- Overproduction of CSF (rare)
  - Choroid plexus papillomas (more common of the two)
  - Diffuse villous hyperplasia of choroid plexus

- Extraventricular (not between ventricles and subarachnoid space) obstruction by tumor, hemorrhage, infection, vascular abnormality, or structural abnormality
  - Possible sites: cerebellar subarachnoid space, basal cisterns, tentorial hiatus (Chiari malformation, achondroplasia) cerebral subarachnoid space
Noncommunicating Hydrocephalus

- An intraventricular obstruction by tumor, hemorrhage, infection, vascular abnormality, or structural abnormality
- Most common cause is aqueductal stenosis, often in association with Chiari II
- Common sites of obstruction
  - Lateral ventricle
  - Foramina of Monro
  - Third ventricle
  - Aqueduct
  - Fourth Ventricle
Fetal Diagnosis of Ventriculomegaly

- Primarily by obstetric ultrasound
- Defined as an atrium of a lateral ventricle larger than 11 mm
  - However, ultrasound cannot confirm whether ventriculomegaly is result of hydrocephalus or loss of periventricular brain tissue in which the vacant space is passively filled with CSF
- May be detected as early as latter part of 1st trimester. Around 20-24 weeks, abnormal dilation of ventricles is more clearly detectable.
Postnatal Diagnosis

- Diagnosis of hydrocephalus is made when the ventricles are enlarged in the absence of cerebral atrophy or dysgenesis
- Radiological Modalities
  - Plain film
  - Ultrasound
  - CT
  - MR
Structural characteristics of hydrocephalus

- Dilation of temporal and frontal horns of the lateral ventricles (often first sign)
- Enlargement of anterior or posterior recesses of third ventricle
- Narrowing of mamillopontine distance
- Narrowing of ventricular angle
- Effacement of cortical sulci

= will return to point again
Hydrocephalus v. Cerebral Atrophy

- Enlargement of temporal horns commensurately with the bodies of the lateral ventricles is probably most sensitive and reliable sign in the differentiation of hydrocephalus from atrophy
  - In atrophy, there is less dilatation of the temporal horns than the lateral ventricles bodies
  - If sylvian fissures are enlarged, dilated temporal horns are not reliable sign
- Large or rapidly enlarging head suggests hydrocephalus
- Small or diminishing head circumference suggests atrophy
Ventricular Angle (left) tends to be smaller in hydrocephalus (shown above) than in atrophy. Frontal horn radius (right) tends to be larger in hydrocephalus than in atrophy.

Courtesy of Dr. Nedda Hobbs and Children’s Hospital Boston Film Library
Plain Films and Hydrocephalus

Prior to newer modalities, diagnosis was made by skull films showing:

- split sutures
- disproportionate craniofacial ratio
- bulging of the anterior fontanel
- erosion of the dorsum sellae
- “hammered silver” appearance of calvarium
Ultrasound and Hydrocephalus

- If anterior fontanelle is open, intracranial structures including ventricles, parenchyma, and vessels are readily visualized in the coronal and sagittal planes.

Used to evaluate for ventricular size, parenchymal and intraventricular hemorrhage, extracerebral fluid collections, cystic lesions, and solid parenchymal masses.

Anterior to posterior coronal images of the patient at age 10 days (study done to look for intraparaenchymal or intraventricular hemorrhage)

Courtesy of Dr. Nedda Hobbs and Children’s Hospital, Boston Film Library
Normal Ultrasound

Anterior coronal image of patient at 10 days

Frontal lobe

Superior aspect of orbits

Courtesy of Dr. Nedda Hobbs and Children’s Hospital, Boston Film Library
Normal Ultrasound

More posterior coronal image of patient at 10 days

Courtesy of Dr. Nedda Hobbs and Children’s Hospital, Boston Film Library
Normal Ultrasound

More posterior coronal image of patient at 10 days

Body of lateral ventricle
Area of thalamus and third ventricle
Corpus callosum
Sylvian fissure

Courtesy of Dr. Nedda Hobbs and Children’s Hospital, Boston Film Library
Normal Ultrasound

Patient at 10 days

Sagittal

Right Parasagittal

Courtesy of Dr. Nedda Hobbs and Children’s Hospital Boston Film Library
Normal Ultrasound

Patient at 10 days

Corpus callosum
Third ventricle
Pons
Sagittal

Thalamus
Fourth ventricle

Courtesy of Dr. Nedda Hobbs and Children’s Hospital, Boston Film Library
Normal Ultrasound

Body of lateral ventricle

Right Parasagittal

Patient at 10 days

Courtesy of Dr. Nedda Hobbs and Children’s Hospital, Boston Film Library
Abnormal Ultrasound

Anterior to posterior coronal images of patient at age 14 months

Courtesy of Dr. Nedda Hobbs and Children’s Hospital Boston Film Library
Abnormal Ultrasound

Anterior coronal image of patient at 14 months

Dilated frontal horns of lateral ventricles

Superior aspect of orbit

Courtesy of Dr. Nedda Hobbs and Children’s Hospital, Boston Film Library
Abnormal Ultrasound

More posterior coronal image of patient at 14 months

Dilated frontal horns of lateral ventricles
Dilated temporal horn of lateral ventricle
Dilated temporal horn of lateral ventricle

Courtesy of Dr. Nedda Hobbs and Children’s Hospital, Boston Film Library
Abnormal Ultrasound

Dilated bodies of lateral ventricles
Dilated temporal horns of lateral ventricles
Dilated third ventricle

More posterior coronal image of patient at 14 months

Courtesy of Dr. Nedda Hobbs and Children’s Hospital, Boston Film Library
Abnormal Ultrasound

Patient at 14 months

Sagittal  Right Parasagittal  Left Parasagittal

Courtesy of Dr. Nedda Hobbs and Children’s Hospital, Boston Film Library
Abnormal Ultrasound

Patient at 14 months

Dilated third ventricle

Dilated body of lateral ventricle

Sagittal

Courtesy of Dr. Nedda Hobbs and Children’s Hospital, Boston Film Library
Abnormal Ultrasound

Patient at 14 months

Right Parasagittal

Dilated frontal horn of lateral ventricle

Dilated temporal horn of lateral ventricle

Dilated body of lateral ventricle

Courtesy of Dr. Nedda Hobbs and Children’s Hospital, Boston Film Library
Comparison of Ultrasound Sections

Coronal images of patient at 10 days

Coronal images of patient at 14 months
Comparison of Ultrasound Sections

Patient at 10 days

Patient at 14 months

Sagittal

Right Parasagittal
CT/MRI Findings in Acute Hydrocephalus

- Temporal horns are preferentially dilated antero-posterially
  - Size > 2 mm
  - Normally, temporal horns are slit-like and barely visible
- Ballooning of frontal horns of lateral ventricles and third ventricle (ie, "Mickey mouse" ventricles)
- Periventricular interstitial edema
- Sylvian and interhemispheric fissures are not visible
CT/MRI Findings in Acute Hydrocephalus (cont’d)

- Fourth ventricle is usually normal in size
- Ratio between largest width of the frontal horns and the internal diameter from inner-table to inner-table at this level should be greater than 0.5
- Ratio of largest width of frontal horns to maximal biparietal diameter > 30%
- Upward bowing of corpus callosum on sagittal MRI
CT/MRI Findings in **Chronic Hydrocephalus**

- Temporal horns may be less prominent than in acute hydrocephalus
- Third ventricle may herniate into sella turcica, which may be eroded
- Corpus callosum may be atrophied (best appreciated on sagittal MRI)
- With long-standing untreated hydrocephalus, white matter will undergo irreversible demyelination
Periventricular Interstitial Edema

- Transependymal CSF resorption from ventricular lumen to the parenchyma
- On CT, appears as hypodensity in periventricular region with indistinct ventricular margins
- On MR, appears as rim of prolonged T1 or T2 relaxation times surrounding lateral ventricles
  - proton density image or a fluid attenuated inversion recovery (FLAIR) image is much more sensitive
- Seen particularly at the superlateral angles of the frontal horns
- Not seen in neonates or young infants (immature brain has normally high water content)
Periventricular Interstitial Edema

T2-weighted image of 5 ½ yo boy who presented with 3 weeks of bifrontal headache, morning vomiting, and blurred vision

Courtesy of Dr. R. Michael Scott and Children’s Hospital, Boston Film Library
Computed Tomography and Hydrocephalus

- **Pros**: widely available, rapid, compatible with life support devices, often requires no patient sedation
- **Should be performed with and without contrast**
Patient at 14 months

Courtesy of Dr. Nedda Hobbs and Children’s Hospital, Boston Film Library
Patient at 14 months

Courtesy of Dr. Nedda Hobbs and Children’s Hospital, Boston Film Library
Expansion of extra-axial CSF spaces

Gray-white matter junction is intact

Note: no evidence of periventricular interstitial edema

Widened sulci with overall effacement of sulci

Enlarged frontal horns + third ventricle = “Mickey Mouse” ventricles

Enlarged temporal horns

Brachycephaly

Patient at 14 months

Courtesy of Dr. Nedda Hobbs and Children’s Hospital, Boston Film Library
Comparison of CTs at Similar Levels of Section

Normal adult

Patient

Patient at 23 months s/p Ventriculoperitoneal (VP) shunt placement

Flattening of posterior occiput

Courtesy of Dr. Nedda Hobbs and Children’s Hospital, Boston Film Library
Patient 2: MRI and Hyrdocephalus

11 yo girl who presented with three days of headache and nausea/vomiting

Courtesy of Dr. R. Michael Scott and Children’s Hospital, Boston Film Library
11 yo girl who presented with three days of headache and nausea/vomiting

Courtesy of Dr. R. Michael Scott and Children’s Hospital Boston, Film Library
Same 11 yo girl with non-enhancing hyperintense mass most consistent with colloid cyst

Courtesy of Dr. R. Michael Scott and Children’s Hospital, Boston Film Library
Comparison of MRIs at Similar Levels of Section

Same 11 yo girl s/p colloid cyst removal one day prior

Postoperative edema in left thalamus

Right pneumocephalus and small subdural hematoma causing mass effect on right frontal lobe
Comparison of MRIs

11 yo girl with colloid cyst

Ventricles are mildly dilated but improved from the prior study.

Before surgery

After surgery

Courtesy of Dr. R. Michael Scott and Children’s Hospital, Boston Film Library
Patient 3: MRI

5 ½ yo boy who presented with 3 weeks of bifrontal headache, morning vomiting, and blurred vision

Courtesy of Dr. R. Michael Scott and Children’s Hospital, Boston Film Library
Comparisons of MRIs

Normal adult  5 ½ yo boy with stenotic aqueduct

Shunts

- Principle of shunting is to establish a communication between the CSF (ventricular or lumbar) and a drainage cavity (peritoneum, right atrium, pleura).
- In principle, a shunt is a plastic tube less than 1/8 of an inch thick that allows one-directional flow of CSF by responding to pressure differences between the ventricle and the cavity to which the shunt terminates.
- There is a valve system that regulates the flow as well as a reservoir, which can be felt through the skin. This reservoir allows for sampling of CSF by needle aspiration.
Shunts

- CSF is simply absorbed in the drainage cavity
- Ventriculoperitoneal (VP) shunt is the most common
  - lateral ventricle is the usual proximal location
  - advantage is that the need to lengthen the catheter with growth may be obviated by using a long peritoneal catheter
- Like all foreign bodies, shunts can malfunction or become infected
- Only about 25% of patients with hydrocephalus are treated successfully without shunt placement

Diagram of VP and VA shunts

From http://www.cinn.org/conditions/hydrocephalus.html
Common Shunt Complications

- **Ventricular end**
  - Blockage
  - Disconnection
  - Migration
  - Hemorrhage
  - Infection
  - Isolated or “trapped” fourth ventricle
  - Secondary craniosynostosis
  - Calvarial thickening
  - Slit ventricle syndrome

- **Atrial end**
  - Thrombosis
  - Infection

- **Peritoneal end**
  - Infection (peritonitis, adhesions)
  - CSF “pseudotumor”, encystment
Patient 4 Shunt Series

Plain films used to aid in confirming proper location of shunt

3 ½ yo boy s/p VP shunt placement

Courtesy of Dr. Ron Becker and Children’s Hospital, Boston Film Library
CSF Shunt Scintigraphy

In different patient evaluated for possible shunt malfunction, there is normal progression of tracer down the shunt catheter, with free spillage into the peritoneal cavity by 15 min. There is no reflux into the ventricles.

Normal transit time ~ 10-20 min. Transit time > 30 min is abnormal.

References

References (cont’d)

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