Hydrocephalus: Teens and Families with Teens

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Jay Riva-Cambrin MD MSc FRCS(C)
Topics

- Hydrocephalus in teens overview
  - Etiologies (underlying causes)
- Endoscopic third ventriculostomy
  - Overview
  - Candidates
  - Relationship with teens
- Slit ventricle syndrome
- Headaches in the shunted teen population
- Precocious puberty in hydrocephalus
- Sports
- Driving
- Transition to adult care
Hydrocephalus in Adolescents

- Hydrocephalus Clinical Research Network (HCRN)
  - Registry – 2006 to present
  - 7 sites
  - Representative of the North American hydrocephalus population
Hydrocephalus in Adolescents

- Only 10% of newly diagnosed pediatric hydrocephalus in children 11-18 years
  - Aqueductal stenosis
  - Intracranial hemorrhage
  - Brain tumors
  - Traumatic brain injury (TBI)

- However, most the 90% diagnosed in infancy or at a young age become teens
2 Populations, 1 Age

- Infants → Teens
  - Chronic stage of disease
  - Maintenance treatment mostly
  - Occasional malfunctions/infections
2 Populations, One age

• Hydrocephalus diagnosed and treated as teen
  – In general, associated diagnosis
    • Ex. Brain tumor

• However, in general, much more amenable for an:
  – Endoscopic Third Ventriculostomy (ETV)
What is an ETV?

- Used instead of a shunt
- Slightly higher upfront risks
- Avoids shunt malfunction and infection
Who are the Best Candidates for an ETV?

- **ETVSS → ETV Success Score**
  - Dr. Kulkarni (Sickkids, Toronto)

<table>
<thead>
<tr>
<th>Score</th>
<th>Age</th>
<th>Etiology</th>
<th>Prior Shunt</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>&lt; 1 month</td>
<td>Postinfectious</td>
<td>Previous shunt</td>
</tr>
<tr>
<td>10</td>
<td>1 month to &lt; 6 month</td>
<td>Myelomeningocele, IVH, non-tectal brain tumor</td>
<td>No previous shunt</td>
</tr>
<tr>
<td>20</td>
<td>6 month to &lt; 1 year</td>
<td>Aqueductal stenosis, tectal tumor, others</td>
<td></td>
</tr>
<tr>
<td>40</td>
<td>1 year to &lt; 10 years</td>
<td></td>
<td></td>
</tr>
<tr>
<td>50</td>
<td>≥ 10 years</td>
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What About Those with Shunts Placed in Childhood and are now in their Teen Years?

- ETV? - technically more difficult

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Slit Ventricle Syndrome

- Shunted ventricles – Much smaller than normal
- Ventricular lining (ependyma) fibrosis – Stiffens
- Brain stiffens – Decrease compliance
- +/- skull arrests growth prematurely
- Shunt blocks → volume quickly changes to pressure
  - Severe pressure symptoms before CT changes (ventricular enlargement)
Slit Ventricle Syndrome

- **RARE**
- More commonly discussed than actually occurs
- Most shunted children have slit-like ventricles
  - But remain compliant
    - Ventricles enlarge
      - obstructed
Thought to Have Had Slit Ventricle Syndrome

• 5 revisions
  – Headaches
  – No CT changes

• Has a true obstruction
  – Headaches
  – Vomiting
  – Drowsy
So Why do Shunted Teens have Headaches if it's Not the SHUNT?

- They just have more headaches than children without shunts
  - Migraines
  - Tension
  - Cluster
  - Unknown
Table 3

Prevalence and Frequency of Headaches for all Study Participants (N=247)

<table>
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<th>Headache within past month:</th>
<th>Statistic</th>
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<tbody>
<tr>
<td>- Yes: n (%)</td>
<td>163 (66.0)</td>
</tr>
<tr>
<td>- No: n (%)</td>
<td>84 (34.0)</td>
</tr>
<tr>
<td>Number of days of headaches in past month:</td>
<td>6.17 (8.0)</td>
</tr>
<tr>
<td>mean (Standard Deviation)</td>
<td></td>
</tr>
<tr>
<td>Number of days of headaches in past month:</td>
<td>3.0 (1.0, 31.0)</td>
</tr>
<tr>
<td>median (minimum, maximum)</td>
<td></td>
</tr>
<tr>
<td>Frequency of days of headaches in past month: n (%)</td>
<td></td>
</tr>
<tr>
<td>- 1 day</td>
<td>48 (29.5)</td>
</tr>
<tr>
<td>- 2 to 15 days</td>
<td>100 (61.4)</td>
</tr>
<tr>
<td>- 16 to 31 days</td>
<td>15 (9.2)</td>
</tr>
</tbody>
</table>
Figure 2. Number of Headache Days Reported by 66% of Children and Adolescents who had Headache over the Last Month (N=163).
• Beginning to recognize that all head problems are not the shunt.

• Headache specialists – After ruled out the shunt.
Puberty

• Puberty starts in:
  – Boys $\rightarrow$ ~11 years old
  – Girls $\rightarrow$ ~ 9-10 years old

• Precocious Puberty (too early)
  – Female under the age of 8 years (7 years)
  – Male under the age of 9 years
Precocious Puberty More Common in Children with Hydrocephalus
Treatment for Central Precocious Puberty

• Treat for:
  – Social issues
  – Short-stature

• Treat if:
  – Girls → puberty < 9y
  – Boys → puberty < 11y

• Treat with:
  – Supprelin (implanted in biceps 1X/year)
  – Lupron Depot-Ped (monthly IM shot)
Sports?

- No restrictions specific to hydrocephalus
  - Discourage but do not prohibit high-impact
    - Football
    - Lacrosse
    - Hockey
    - Wrestling
    - Gymnastics
Driving?

- No restrictions specific to hydrocephalus
- Epilepsy or seizures
  - Common co-morbidity
  - Would be the driving limiting diagnosis
  - Rules/law varies by state
Transition to Adult Care

- Huge and very hot topic in medicine
  - All pediatric chronic disease
  - Session later dedicated to just this topic
Transition to Adult Care

• GENERAL PRINCIPLES:
  – Shunts less problematic in adulthood
    • Unknown?- no longer a moving target?
  – Still need follow-up
    • But less frequent
    • Adult neurosurgeons
      – “call me if you have a problem or a concern”

• Some specialty clinics are emerging